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Insight in Progressive Supranuclear Palsy (PSP): A Study of Intra-Individual Awareness

MSc HCN Dissertation

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Abstract

Deficits in awareness challenge patient's health and safety and can decrease not only the patient's quality of life but also that of the people around them (Rymer et al., 2002)

Despite the frequency of awareness deficits in neurological diseases and its clinical relevance, the study of awareness in neurological conditions is still not fully understood. Moreover, it is not clear whether impaired awareness is a unitary concept or it can dissociate across different functional domains (Vasterling, Seltzer, Foss and Vanderbrook 1995). Due to the frontal pathology present in Progressive Supranuclear Palsy (PSP), many patients suffer from awareness deficits. However, the study of awareness in PSP has only recently been studied (O'Keeffe, et al., 2007). Following O'Keeffe et al's (2007) multidimensional approach to awareness, the present study examines differences in awareness types (metacognitive, anticipatory and emergent awareness), the specificity of awareness deficits across cognitive domains and the effects of mood on awareness estimations across PSP patients, their primary carers and a group of controls. Results show that PSP patients have a specific pattern of awareness with not only general differences between emergent, anticipatory and metacognitive awareness (emergent being better than anticipatory and metacognitive awareness), but also domain-specific differences across different cognitive areas (with patients making more mistakes in executive tests than in any other types of tests). Moreover, the study also shows that the estimations made by the primary carers on the patients' as well as on their own performance are inaccurate.

Keywords: Progressive Supranuclear Palsy, subcortical dementia, awareness, specificity.

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1. Introduction

Progressive Supranuclear Palsy (PSP), also known as the Steele-Richardson-Olszewski syndrome (SRO), was first described by Steele, Richardson and Olszewski (1964) as a late-onset progressive neurodegenerative disease, with ocular, motor and mental features (Steele, Richardson and Olszewski, 1964).

This disease, together with corticobasal degeneration (CBD) and multiple system atrophy (MSA), is often classified as an 'atypical parkinsonian syndrome' or 'Parkinson-Plus syndrome', due to the characteristic akinetic-rigid syndrome which is typical of Parkinson's disease (PD) (Bak, 2007, p. 80). However, despite some of the similarities in symptomatology that PSP and PD patients share, PSP is not a variant of PD but instead an entity in its own right, with specific neuropathological and neuropsychological characteristics.

The terms 'atypical parkinsonian syndrome' or 'Parkinson-Plus syndrome' which are often used to refer to this disease, suggest that PSP is mainly a movement disorder. However, neuropathological and clinical studies have shown that PSP patients present with a certain degree of frontal pathology which results in cognitive decline.

The impairment of functions within the frontal lobes can be very varied and consequently patients can experience a wide range of deficits. This has caused a lot of debate as to the degree to which the frontal functions can be divided.

This study however focuses on one particular frontal function: the awareness patients have of their own impairments. The study will first describe the neuropathology and neuropsychology of PSP and will then examine the phenomenon of insight, and how it is impaired in PSP patients.

1.1 Neuropathology

The original publication by Steele, Richardson and Olszewski in 1964 provided the first detailed histological description of the neuropathology of PSP and started a wave of research trying to explain the neuropathological basis of PSP and its progression.

PSP is currently classified among the tauopathies, a group of disorders which also include Alzheimer's disease (AD), Pick's disease (PiD), corticobasal degeneration (CBD), the NFT predominant form of senile dementia (NFT-SD), argyrophilic grain disease (AGD), and parkinsonism dementia complex of Guam (Guam PDC) (Armstrong, Lantos and Cairns, 2007). These diseases are classified as tauopathies due to abnormally phosphorylated tau-protein present in neurons and glia in subcortical and cortical structures (Ahmed, Josephs, Gonzalez, DelleDonne and Dickson 2008). In addition to the changes in the tau-protein, degenerative changes in PSP include neuronal loss, gliosis, neurofibrillary tangles (NFT), granulovacuolar change (GVC), and demyelination, mainly affecting the globus pallidus, subthalamic nucleus, subtantia nigra, dentate nucleus of the cerebellum, and brain stem tegmentum (Daniel, de Bruin and Lees, 1995; Armstrong, et al., 2007). The current neuropathological consensus for PSP has been proposed by the National Institute of Neurological Disorders and Stroke (NINDS) and requires a high density of NFT in at least three of the following sites: the subthalamic nucleus, the pallidum, the subtsantia nigra or the pons. The criterion also requires a low to high density in at least three of the following sites: the oculomotor complex, the striatum, the medulla or the dentate nucleus with neuronal loss and gliosis are variable and amyloid deposits and neuritic plaques are notably absent (Hauw, et al., 1994).

1.2 Clinical Diagnosis

The clinical symptoms in PSP mirror the neuropathology of this disease (see Table 1 for the clinical diagnosis of PSP).

The common initial symptoms of PSP are usually fatigue, lethargy and a feeling of weakness. Some patients experience a general slowness in bodily movement, or changes in personality including depression and apathy. Diplopia and other symptoms of visual dysfunction are also common in early manifestations of PSP (Litvan and Agid, 1992, p.20). These symptoms may be manifested in patients' driving, judgement of distances and falling. Gait is also impaired and patients tend to use short steps, shuffle, and 'freeze' in place. Moreover, many patients experience sudden falls, usually backwards which occur without any warning signs, increasing the patients' feeling of unsteadiness and fear of falling (Litvan and Agid, 1992, p. 20).

With regard to their visual abilities, fixation instability, which is manifested in macro square-wave jerks, defective visual suppression of the vestibuloocular reflex, and loss of opticokinetic nystagmus are the most common early abnormalities present in PSP patients, preceding overt gaze palsy.

Patients with this disease seem to also present with reduced spontaneous blinking, retraction of the eyelid (Cowper's sign), 'apraxia' of eyelid opening and closing (i.e. supranuclear paresis of eyelid opening and closing) and facial hypomimia, which can account for the 'blank staring' facial expression with an appearance of surprise common in PSP patients (Litvan and Agid, 1992, p. 24).

Early in the course of the illness, patients may experience dysarthria and dysphagia, which tend to be disproportionate to the severity of the parkinsonism, and

slowness of the tongue and mouth movements (Litvan and Agid, 1992, p. 24). These can cause patients to lose weight and to have to reduce their diet to smooth, soft food.

In relation to speech, patients may have slow, slurred and monotonal speech and lose any facial expression or expressive gestures to accompany the speech. As to their posture, it is usually erect, with a slight hyperextension of the trunk (Litvan and Agid, 1992, p. 26).

Finally, PSP patients can have some cognitive impairments such as forgetfulness, slowness of thought processes and frontal lobe dysfunction, which present with the absence of true apraxia, aphasia or agnosia. In depth study into the neuropsychology of PSP will be provided in the following sections.

Table 1. Clinical diagnostic criteria for PSP

Essential for the diagnosis
Onset over age 40
Progressive course
Bilateral supranuclear disorder of ocular motility
Hesitance of voluntary down-gaze
Impaired vertical optokinetic nystagmus
Poor suppression of vertical vestibuloocular reflex
Rigidity with axial predominance
Bradykinesia
No evidence of other diseases that could explain the above features
Confirmatory manifestations
Poor or absent response to levodopa therapy
Severe bradyphrenia with frontal lobe features (grasping, perseveration, utilisation)
Axial dystonia with cervical hyperextension
Gait impairment and frequent falls, postural instability
Dysarthria and dysphagia
Ocular fixation instability with macro square-wave jerks
Apraxias of eyelid opening or closing, extreme infrequency of eyeblink
Echolalia, palilalia

Note. Table taken from Litvan and Agid (1992), p. 18.

1.3 Differential Diagnosis

Given the diffuse pattern of degeneration present in PSP and the fact that PSP and Parkinson's disease (PD) share some common pathology, it is not surprising that PSP is usually classified as an atypical parkinsonian syndrome or even occasionally misdiagnosed as being PD (Fisk, Goodale, Burkhart and Barnett, 1982). The main differences however, are related to the presentation of the symptoms and the responsiveness to medication. Early PSP has a more symmetrical presentation of symptoms than early PD and whilst early PD is generally responsive to L-dopa

medication, PSP is not (Joseph and Young, 1999, p. 229). It has been suggested that the extensive damage to the basal ganglia output pathways present in PSP patients is responsible for the L-dopa unresponsiveness as well as the axial parkinsonism which some patients present, whilst the destruction of the midbrain reticular formation is responsible for the supranuclear gaze palsy (Hauw et al., 1994). Furthermore, PSP usually has a faster progression, with most patients succumbing within six to seven years from the onset of symptoms (Purcell and Reich, 1997; Rehman, 2000).

Progressive supranuclear palsy also shares some common neuropathology with Alzheimer's disease (AD), and as mentioned above, they can sometimes be misdiagnosed as one another. However, neuropathologically it is possible to distinguish between PSP and AD due to differences in the type of tau protein deposition, topography and amyloid deposition present in each of the diseases (Morris et al., 2002).

It is also possible to distinguish between AD and PSP from a neuropsychological perspective. As Albert and colleagues (1974) suggested, the cognitive and behavioural changes observed in PSP patients, are more indicative of what they called a "subcortical dementia" than of those observed in "cortical dementias" such as AD (Albert, Feldman and Willis, 1974).

1.4 Cognitive and Behavioural Impairments

In their seminal article, Steele et al. (1964) described nine cases with PSP, seven of which also presented intellectual impairments, six had personality changes such as irritability, suspiciousness or untidiness, two cases were described as presenting with emotional liability and finally one case presented with signs of depression. It is therefore clear that cognitive and behavioural impairments are a fundamental part of the PSP diagnosis.

It is difficult however to determine the frequency and severity of dementia in PSP. This is mainly due to the fact that many studies have used inappropriate tests to determine the presence of dementia in PSP patients. For instance, the use of the Mini Mental State Examination (MMSE) to determine the presence and characteristics of dementia in PSP relies heavily on intact language abilities and is insensitive to frontal lobe deficits (Bak, et al., 2005a). This can be a problem for the diagnosis of PSP since many patients are unable to speak, read or write. In addition, several studies on PSP patients have reported some degree of cortical pathology, especially in the frontal lobes (Bak and Hodges, 1998; Bak, Crawford, Hearn, Mathuranath, and Hodges, 2005b). This means that tests such as MMSE will not be useful in detecting this type of disease, and thus studies relying on these types of tests will have underestimated the frequency and type of dementia in this disease.

Currently, the cognitive and behavioural symptoms in PSP patients are mainly disturbances of thought, executive dysfunction, and some behavioural disturbances such as apathy, disinhibition, depression, and anxiety (Rehman, 2000). Executive functions are those involved in the realisation of goal-directed behaviour, which can be expressed through either a motor or a mental act (Litvan and Agid, 1992, p. 228) and studies have shown these functions to be impaired after damage to the frontal lobes (Shallice and Burgess, 1991). Given the extensive damage to subcortical structures and the frontal behavioural symptoms that PSP patients show, it therefore seems reasonable to explain the cognitive and behavioural symptoms as due to dysfunction of a ‘subcorticofrontal system’. In fact, Albert et al (1974) suggested the term ‘subcortical dementia’ in order to describe these behavioural and cognitive symptoms, and suggested they were caused by deactivation of the cerebral cortex due to lesions to subcortical structures (Litvan

and Agid, 1992, p. 223). Subcortical dementia as defined by Albert and colleagues (1974), presents with symptoms associated with dysfunction of the prefrontal cortex as well as the neuropathological involvement of subcortical structures but without the presence of major cortical lesions (Albert et al., 1974). An imaging study by D'Antona et al (1985) used positron tomography and established a correspondence between the clinical signs suggesting a prefrontal lobe dysfunction in PSP patients and the presence of cortical hypometabolism affecting mainly both frontal lobes, which the authors suggested resulted from subcortical lesions. With this finding, D'Antona and colleagues (1985) strengthened the concept of subcortical dementia and confirmed the presence of frontal lobe dysfunction in PSP.

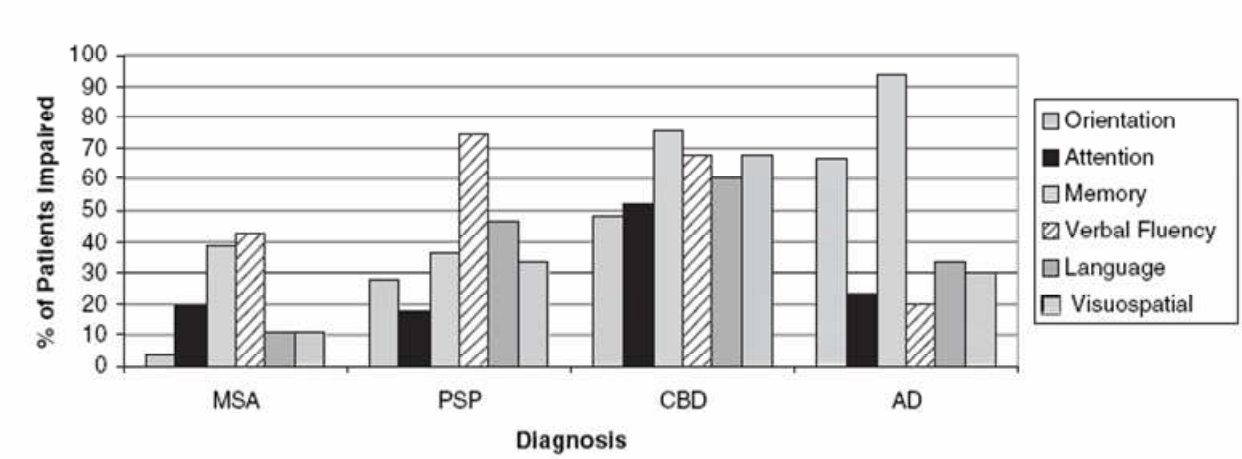
In an attempt to distinguish between cortical and subcortical dementia, Cummings and Benson (1984) classified patients' impaired functions into either 'instrumental' or 'fundamental'. Patients with cortical dementia such as AD, are impaired in 'instrumental' functions such as memory, language or praxis; whilst patients with subcortical dementia, such as PSP, are impaired in 'fundamental' functions such as timing, attention, motivation or programming. In accordance to this distinction Lhermitte, Pillon and Serdaru (1986) pointed out that PSP patients tend to be abnormally dependent on stimulations by the examiner and seem unable of self-guided behaviour or giving their own answers. Moreover, they are unable to inhibit the tendency to use objects presented in front of them and to stop some motor responses such as clapping their hands, once the action has been started. Clinical observation supports this distinction.

Albert and colleagues' paper (1974) started an important theoretical debate concerning the role of subcortical structures in cognition and led to an increase in the number of studies on the cognition of diseases with subcortical pathology such as PSP.

Several studies in the 80s attempted to delineate the specific cognitive deficits in PSP and other atypical parkinsonian syndromes. In 1986, Pillon, Dubois, Lhermitte and Agid compared three groups of patients - namely PSP, PD and AD patients- to a matched control group and found that the three groups of patients had similar results on verbal, visuospatial and global memory tests. However, PSP patients showed significant deficits on tests of attention and executive tasks. Three years later, Milberg and Albert (1989) compared the performance in a range of cognitive tests of a group of PSP patients to a group of AD patients and found that whilst AD patients showed verbal and non-verbal memory impairments, the PSP group seemed to be impaired in the lexical fluency test, supporting the idea of executive dysfunction in PSP patients due to frontal lobe dysfunction.

More recently, Bak and colleagues (2005b) compared the performance of three groups of patients thought to have subcortical dementia (PSP, CBD and multiple system atrophy (MSA)), with a group of AD patients, thought to have a more cortical dementia. Their study showed that PSP, CBD and MSA patients were particularly impaired on the verbal fluency subtest of the Adenbrook's Cognitive Examination (ACE). The authors suggested this was due to damage to the basal ganglia, which was present in all three diseases, and which corresponded to the subcortical core deficit (Bak, et al., 2005b). PSP and, especially, CBD showed deficits in other cognitive areas as well (*see* Figure 1) , which Bak et al. (2005b) suggested reflected the extensive frontal and fronto-parietal damage present in those patients (Bak, et al., 2005b).

Figure 1. Pattern of impairment in ACE Sub-tests in four patient groups (MSA, PSP, CBD and AD (Bak et al., 2005b).



Note. Figure taken from Bak, et al. (2005b). MSA= Multiple system atrophy, PSP= Progressive supranuclear palsy, CBD= Corticobasal degeneration, and AD= Alzheimer's disease.

In addition to these cognitive impairments, patients with PSP manifest behavioural disorders that resemble those encountered in patients with frontal lobe lesions (Lhermitte et al., 1986) such as bluntness of affective expression, depression, or less often, an inappropriate laughing or crying, euphoria, paranoia, irritability and even occasional outbursts of rage (Albert, et al., 1974). These symptoms can interfere with the cognitive assessment and cause additional problems at the time of interpreting the results. Moreover, patients' lack of insight and persistence of emotions can also increase the difficulty of neuropsychological assessment making it extremely difficult for the examiner to delineate the cognitive impairments in PSP patients.

1.4.1 Insight and insight specificty

Despite the cognitive and motor impairments present in PSP, some patients appear to be unaware of their deficits; they seem to suffer from what Babinski (1914) termed 'anosognosia'. Anosognosia refers to one's inability to recognise one's deficits

(McGlynn and Schacter, 1989) and is generally related to frontal lobe dysfunction (Michon, Deweer, Pillon, Agid and Dubois, 1994), such as that found in PSP.

The study of awareness is of both theoretical and clinical interest and has been the subject of extensive investigations mainly in patients with FTD and AD. It is important to study insight since unawareness of one's deficits has been linked to negative aspects of disease outcome such as increased stress and caregiver burden (Seltzer, Vasterling, Yoder and Thompson, 1997), poor patient-caregiver relationship (Hutchinson, Leger-Krall, and Wilson, 1997), poor medication compliance and the performance of risky and life-threatening activities (McGlynn and Shacter, 1989; Cotrell and Wild, 1999).

Moreover, *ad hoc* studies using clinician ratings of awareness of deficits in AD patients have been shown to have a great influence on the outcome of psychosocial and neuropsychological rehabilitation (Koltai, Welsh-Bohmer and Schmechel, 2001). This suggests that an understanding of awareness and the use of appropriate measures to test awareness of deficits is of great importance and it can assist in the selection of appropriate interventions for individuals suffering from anosognosia.

Awareness is a very complex concept, consequently studies exploring awareness in dementia seem to use terms that relate to different concepts (Markova, Clare, Wang, Romero and Kenny, 2005). It is often used synonymously with consciousness, in the sense of being aware of incoming information, or processing certain knowledge, thoughts or intentions. However, awareness is more than just consciousness; the concept of awareness also includes self-consciousness and self-knowledge (Clare, 2004).

Moreover, the complexity of the concept of insight increases when you take into account the 'object of insight'. Markova and Berrios (2001) claim insight can only be

understood in relation to something; in this case the 'object of insight'. For instance, regarding this study, the 'object of insight' refers to a particular mental or physical state in relation to which insight can be assessed (Markova and Berrios, 2001).

Therefore, for the purposes of this study it is essential to make clear that insight is defined as a form of self-knowledge patients hold concerning an illness or impairment that affects them, which includes not only information about the particular pathological state, but also how this state affects them and their interaction with their environment. It is also important to distinguish anosognosia from other terms such as *anosodiaphoria*, which refers to indifference or a diminished emotional concern to any type of neurological impairment (McGlynn and Shacter, 1989).

Disturbances of awareness have been documented in a wide range of neurological disorders. There are many ways in which patients can be unaware of their deficits; for instance a person may demonstrate lack of awareness by actively denying any difficulty or acknowledging a degree of difficulty but trying to give an explanation for it, or even failing to acknowledge there is a problem at all (Khilstrom and Tobias, 1991).

Stuss and colleagues (2001) classified awareness disturbances into four types: (a) pervasive disturbances of consciousness such as those found in vegetative states (Zeman, 1998), (b) domain-specific unawareness, or unawareness of the impairment of a particular function such as those found in patients with hemiplegia, hemianopia, visual agnosia or prosopagnosia (Bisiach, Vallar, Perani, Papagno and Berti, 1986) (c) executive unawareness (i.e. the impaired ability to regulate one's own behaviour) and (d) impaired self-awareness (i.e. the inadequate evaluation of one's impairments and possible consequences).

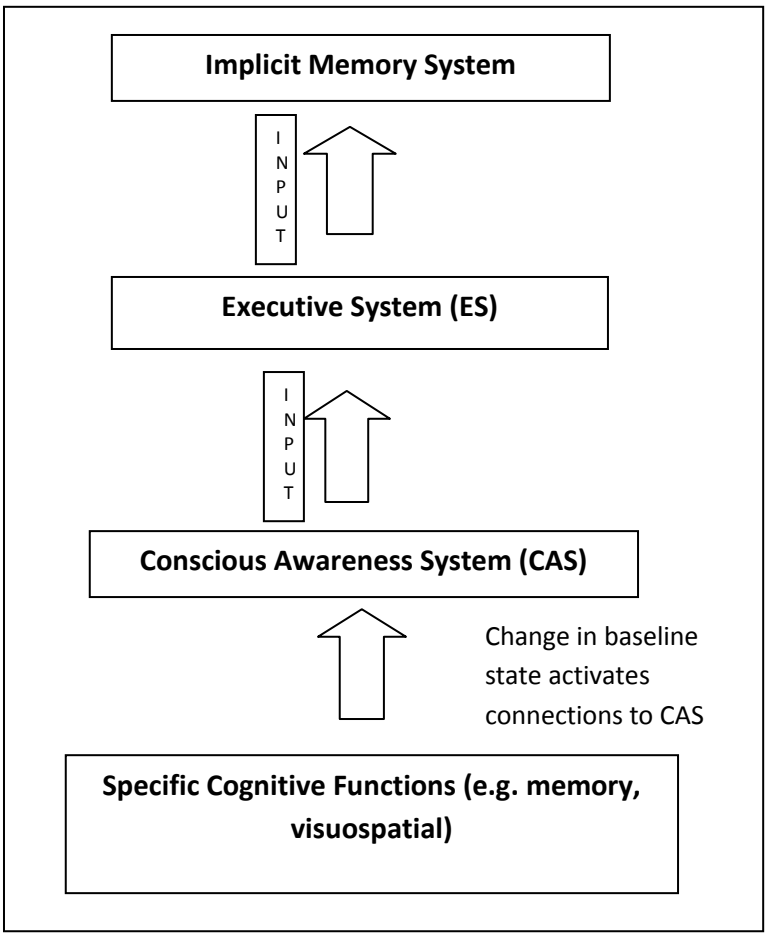
These diverse manifestations of unawareness that can be observed in different neurological disorders have stimulated the development of several neuropsychological, anatomical and psychiatric models of awareness (Clare, 2004).

However, it seems that despite the psychometric view of awareness as a unitary construct; it is instead a multidimensional complex which encompasses different levels and which therefore needs a complex model to explain the variations of awareness seen in different neurological disorders.

Based on neuroanatomical and clinical evidence suggesting the involvement of the frontal lobes in executive functions, Fernandez-Duque and Black (2007) and Stuss et al (2001) claimed that damage to the frontal lobes was implicated in disturbances of executive and self-awareness. Executive control includes a variety of functions, such as monitoring one's recent and past cognitive performances, establishing future goals, inhibiting over learned responses as well as altering behavioural patterns in response to feedback. It therefore seems reasonable to predict that damage to any of these executive 'sub-functions' will produce different states of awareness in a patient (Souchay, Isingrini, Pillon and Gil, 2003). For instance, Nelson and Narens (1990) suggested the term 'metacognition' to refer to the knowledge people have about a certain task, the strategies used, or even about their own cognitive abilities. They claimed that metacognition, *i.e.* awareness, included two main functions: one responsible for monitoring the collection of information received about one's own performance, and another function responsible for controlling one's own behaviour. These abilities fit perfectly with the functions that Souchay and colleagues (2003) suggested were part of the executive function system. This basic model of awareness, however, cannot explain all the different types of awareness deficits that patients can exhibit.

A more complete model of awareness is that offered by the Dissociable Interactions and Conscious Experience (DICE) model (Shacter, 1990) (*See* Fig. 2) and its later development to the Conscious Awareness Model (CAM) (Litvan et al., 1997) both of which were devised to explain awareness deficits in AD.

Figure 2. The Dissociable Interactions and Conscious Experience (DICE) model of awareness (Shacter, 1990)



The lowest level of this model contains modules which represent specific cognitive functions. When a change in the baseline resting state occurs, a module and its links to the Conscious Awareness System (CAS) activate. This results in conscious awareness of information being processed. The CAS then inputs to the Executive

System (ES), responsible for synthesising the information needed for complex functions, and in turn provides input to the Implicit Memory System (IMS) (Clare, 2004).

Damage could occur at any of the multiple levels and manifest itself as a different form of awareness deficit. For instance, individual modules could be selectively disconnected from CAS meaning that information would thus not reach conscious awareness, thus causing domain-specific unawareness. Furthermore, damage to the CAS would result in the output of inappropriate responses and hence cause a general awareness deficit of basic cognitive impairments across domains. Finally, damage to the ES would cause impairments in the synthesis of information arriving from the CAS and as a result cause awareness deficits of complex functions mediated by ES (Clare, 2004).

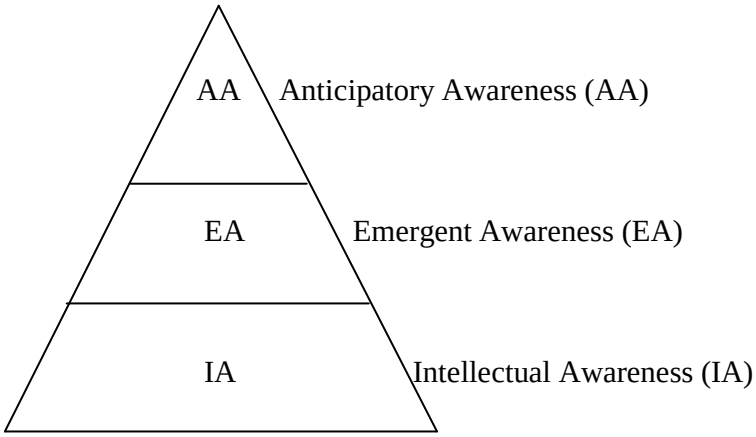
However, there are several limitations to this model; for instance, it fails to account for the relationship observed between loss of insight and right hemisphere damage (McGlynn and Shacter, 1989). In addition, the model does not account for psychological or psychosocial factors and it also fails to explain how memory deficits, such as those seen in AD, are involved in metacognition or in the mechanisms available for metacognitive output.

Stuss and colleagues (2001) proposed yet another cognitive neuropsychological model of awareness which accounted for the clinical observation of different categories of awareness disturbances in neurological disorders. In their model, the brain receives information from the world and then engages in a modelling process in which it constructs a model to fit this information. The model is constantly being updated by reality checking systems or feedback (Clare, 2004). The modelling process

encompasses four levels; the basic level mediates arousal, the second level is in charge of the perceptual analysis and the engaging of complex motor activities, the third level controls executive functions and goal-directed behaviour, and the final level corresponds to the current state of consciousness (Clare, 2004). At each level, a model is produced and compared to incoming information. Higher levels use top-down processes to control lower levels, whilst bottom-up processes are used by lower levels to activate modelling at higher levels (Clare, 2004). Therefore, damage to any of the levels can produce different manifestations of awareness (Stuss et al., 2001). However, as with the DICE, a significant limitation of this model is the fact that it does not account for any psychological or psychosocial factors.

In addition to the neuropsychological and anatomical models of awareness, clinical models have also been developed in order to guide the rehabilitation process and understand the heterogeneity of insight deficits. An important clinical model of awareness based on data from head-injury patients was proposed in 1989 by Crosson and colleagues (*see* Fig. 3). This hierarchical model of awareness divided awareness into three interdependent types: intellectual awareness (IA) (i.e. the patients' own ability to recognise their impairments), emergent awareness (EA) (i.e. the patients' ability to recognise difficulties as they appear), and finally, anticipatory awareness (AA) which is responsible for the patients' ability to predict any difficulties due to their deficits.

Figure 3. Crosson et al. (1989) hierarchical model of awareness



Note. Awareness represented as a pyramid. IA is the foundation for EA and AA. Some degree of EA is necessary for AA.

Toglia and Kirk (2000) proposed a more interactive model of awareness in which they differentiated between 'metacognitive' knowledge similar to intellectual awareness, and 'online monitoring' of performance during tasks, which incorporated elements of both emergent and anticipatory awareness.

Evidence for these models comes from studies in which associations have been found between impaired insight in AD or frontotemporal dementia (FTD) patients and the presence of dysexecutive symptoms (Weinstein, Friedland and Wagner, 1992; Dalla Barba, Parlato, Iavarona and Boller, 1995; Souchay, Isingrini, Pillon and Gil, 2003). This is because each of these models involves an executive system which is responsible for updating and comparing information from the real world with stored information. Damage to this system will produce disturbances of awareness.

Reed, Jagoust and Coulter (1993) found a correlation between hypoperfusion of the right dorsolateral frontal lobe and awareness deficits in AD. Consistent with these results, a study by McDaniel and colleagues (1995) proposed an association between frontal cortex pathology and awareness deficits (McDaniel, Edland and Heyman, 1995).

Neuroimaging studies have given further support for the involvement of the frontal cortex on awareness of deficits. For instance, a study by Mendez and Shapira (2005) investigating the association between insight loss and frontal lobe degeneration in FTD using fMRI, found that loss of insight was greatest in patients with right hemispheric hypoperfusion or hypometabolism, especially in the frontal lobes.

It seems, however, that most studies have focused on distinguishing between different types of awareness deficits in patients (*e.g.* metacognitive, anticipatory or emergent) but have not taken into account the object of insight. The extent to which impaired awareness is unitary or it is different depending on the functional domain is still unknown. Green and colleagues (1993) provided preliminary evidence that dissociations across functional domains could be possible in patients with AD. Their study found that patients and carers reported varying levels of awareness depending on the domain they were testing (remote or recent memory, attention and everyday activities) (Green, Goldstein, Sirockman and Green, 1993). In fact, a study two years later by Vasterling and colleagues (1995) found adissociation in awareness impairments in a group of AD patients, between what they called '*higher-order'* information and integration functions, such as memory and self-care, and '*lower-order'* functions. Moreover, their results revealed that these higher-order functions were the first functions to be impaired, and were affected by factors such as disease stage and main area of cognitive deficit. Nevertheless, insight specificity in dementia patients remains largely unknown and more information is needed, on other patients rather than AD, to understand this phenomenon more clearly.

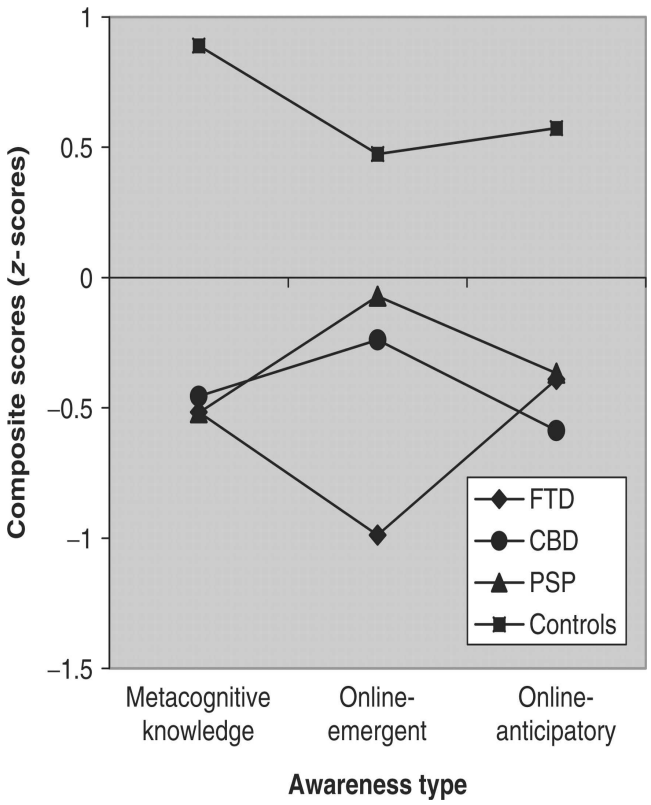
Despite extensive frontal lobe damage in PSP patients (Cordato, Duggins, Halliday, Morris and Pantelis, 2002), insight, was only recently studied in this disease.

O'Keeffe and colleagues (2007) carried out a multi-dimensional study to insight in which they used the clinical models of awareness proposed by Crosson and colleagues (1989) and Toglia and Kirk (2000) across three patient groups, namely PSP, CBD and FTD patients. They studied three dimensions: metacognitive awareness (knowledge of one's abilities), online emergent awareness (online monitoring of errors), and online anticipatory awareness (predictions of one's abilities).

Anticipatory awareness was measured by asking participants to rate what they thought their performance would be like in a series of cognitive tests. Emergent awareness was examined using the Sustained Attention and Response Task (SART) by asking participants to indicate every time they thought they had committed an error. Finally, metacognitive awareness was measured by asking them a series of questions about their abilities and what they thought others' abilities were.

O'Keefe et al (2007) found that insight in PSP patients was impaired, especially their metacognitive and anticipatory awareness (Fig. 4).

Figure 4. Awareness deficits in PSP, CBD and FTD patients (O'Keefe et al., 2007)



Note. FTD= frontotemporal dementia; PSP= progressive supranuclear palsy; CBD= corticobasal degeneration.

However, despite the cognitive deficits being relatively specific in PSP (Bak et al., 2005b), O'Keeffe et al. (2007) did not distinguish whether awareness was task-specific. The present study aims to look at intra-individual awareness in patients and their caregivers as well as the awareness specificity and the influence of depression and anxiety on performance estimations.

1.4.2 Depression and its relationship to insight

Neurological diseases associated with changes in mood, especially depressive episodes, are very common (Stuss and Knight, 2002, p. 378). Depressive episodes can be described as presence of a persistent negative mood state as well as attention, motivation, motor and mental speed and appetite disturbances (American Psychiatric Association, 1994).

Some authors have proposed a link between depression and preserved insight in dementia patients, however studies examining this association have yield inconclusive results. It is important to study this relationship since it can help with the understanding of the dementia and can have very important implications for the choice of therapeutic strategies (Verhey, Rozendaal, Ponds and Jolles, 1993).

On the one hand, some studies (Sevush and Leve, 1993; Feher, Mahurin, Inbody, Crook and Pirozzolo, 1991) have found that the more depressed patients with dementia are, the more aware of their impairments they are. A study by Smith and colleagues (2000) studied the role of depressive symptoms in mediating impaired insight in a group of AD patients and they found that AD patients with higher levels of depressive symptoms also demonstrated greater awareness of their deficits. This study highlights the importance of accounting for depression in any study on awareness in patients with dementia.

A possible explanation for the results pointing to a relationship between depression and awareness of deficits in dementia patients was proposed by Sevush and Leve (1993) and Feher and colleagues (1991). They suggested that depression in patients with dementia might have developed as a reaction to the awareness patients have of their impairments and disease progression.

However, on the other hand, Reed and colleagues (1993) studied the relationship between major depressive symptoms in a group of patients with AD and awareness deficits and found there was no association between them. Consistent with these results, De Bettignies, Mahurin and Pirozzolo (1990) and Verhey et al (1993) used the Hamilton Depression Rating Scale (HDRS) on a group of mixed patients and found no association.

Affective and behavioural changes are common in PSP (Chiu, 1995). The most commonly reported symptoms are personality changes and an increased irritability, impulsivity, untidiness, suspiciousness, emotional liability, apathy and even euphoria. However, depression can also be present in many patients, but whether the depressive symptoms present in PSP patients are part of a depressive illness or whether they are part of a subcortical dementia remains unclear (Chiu, 1995). It is likely that the number of PSP patients with depression is overestimated due to the fact that many studies report 'slowing of mental processes', 'inability to concentrate' or 'slowing of movements' as characteristic depressive symptoms in PSP, whilst these symptoms might just be features of PSP per se (Chiu, 1995).

A study by O'Keeffe and colleagues (2007) examining awareness deficits in PSP as well as in FTD and CBD, found that patients who scored highly on the Hospital Anxiety and Depression Scale (HADS) presented with poor anticipatory awareness, i.e. they performed poorly when asked to predict what they thought their performance on a task would be. This suggests that preserved insight may be linked with lower emotional dysfunction in PSP patients. This issue will be studied further in the present study.

1.5 Present Study

The aim of this study is threefold. Firstly, it examines the accuracy of PSP patients' and their caregivers' judgements of their performance on several cognitive tests prospectively (before doing the test) and retrospectively (after test). Secondly, it explores whether PSP patients and their main caregivers are aware of the specificity of their cognitive impairments. Finally, the study examines whether patients' and caregivers' performance estimations' are affected by depression or anxiety.

- *Prospective and retrospective performance estimations*

A described by Green and colleagues (1993), awareness of deficits is defined in this study as the degree to which estimations made by the participants agree with their actual performance. Following O'Keeffe et al. (2007) study on awareness deficits in PSP and using Crosson et al (1989) and Toglia and Kirk's (2000) multidimensional model to awareness deficits, this study aims to replicate O'Keeffe et al's (2007) findings that PSP patients exhibit poor insight in comparison to controls, especially in the areas of metacognitive and anticipatory awareness. The present study however, will use a different neuropsychological battery to test performance and awareness. The battery includes an array of hard and easy tasks as well as questions to predict their performance before and after reach test. Finally, this study will compare the insight rating of a group of PSP patients to that of their carers and a control group of volunteers.

- *Insight Specificity*

Since PSP patients have been found to be more impaired in executive functions (e.g. verbal fluency) than in visuo-spatial tasks (Bak et al., 2005b), they are expected to perform worst at executive function tasks than at visuospatial tasks. Based on previous

research and neuropsychological models of awareness that emphasize potential functionally specific dissociations, we predicted that insight accuracy would vary depending on the cognitive domain assessed. Thus, this study will reveal whether patients are able to differentiate their impaired performance in executive tasks from their relatively normal performance on visuospatial tasks. A good estimation of these tasks will mean that patients do not have a generalised awareness deficit, but instead they are impaired on specific aspects of insight.

- *Mood*

Some studies have found a significant relationship between depression and insight. Sevush and Leve (1993) found a relationship between the level of depression and insight in dementia patients and concluded that depression might have developed as a reaction to the awareness of the disease by the individuals. This question investigates more in depth the proposal by O'Keeffe et. al (2007) that preserved insight is associated with lower emotional dysfunction.

We predict that patient's whose main problem is insight, will have a tendency to overestimate their performance, whereas if depression is the main problem, they will tend to underestimate their performance.

2. Materials and methods

2.1 Participants

Eligible participants who agreed to participate gave informed consent according to the Edinburgh University Ethics Committee.

A total of thirty individuals took part in this study. This included 10 patients and their respective 10 caregivers and 10 neurologically healthy controls.

2.1.1 PSP patients (n=10) and caregivers (n=10)

Patients were recruited from the PSP Association Europe throughout the United Kingdom (Edinburgh, Glasgow, Northampton, Liverpool and Newcastle) from March 2008 to July 2008. All PSP patients had been referred to the PSP Association Europe by specialist neurologists. Patients were 7 men and 3 women whose ages ranged from 52 to 81 and who had had a mean of 12.9 many years of full-time education (*see* Table 2).

Table 2. Descriptive statistics for gender age and education for the three participant groups

			Age (years)		Full time education (years)	
Group	n	m:f	M (SD)	Range	M(SD)	Range
PSP patients	10*	7:3	66.8(8.6)	52-81	12.9(3.2)	9-18
Caregivers	10	3:7	63.9(12.7)	48-83	12.8(2.8)	10-17
Controls	10	5:5	70.2(7.5)	55-78	15.4(3.2)	10-18

Note. m= males; f= females; M = mean; SD= Standard deviation; PSP = Progressive supranuclear palsy. There were no significant differences between demographic variables for controls, carers and PSP patients.

Most patients (n= 9) had first been diagnosed with PSP at least 1 year ago, whilst 1 patient had been diagnosed more recently, 2 months ago. All caregivers and patients however believed patients had suffered from PSP well before having been first

diagnosed. All patients were taking some form of medication. Two patients were taking Levodopa medication, and three patients were taking anti-depressant medication.

All main caregivers were the patients' spouses (7 women and 3 men). The caregivers' ages ranged from 48 to 83, and had had an average of 12.70 years of full-time education. Nine caregivers were retired and cared for their partners' full time, whilst one still kept his job and thus had additional help during the day.

One caregiver was on anti-depressant medication and two were taking medication for high blood pressure and cholesterol.

2.1.2 Controls (n=10)

Participants in this group (5 women and 5 men) were recruited through the volunteer panel of the Psychology department of the University of Edinburgh. Control participants were sent a letter with information about the study and a prepaid envelope with a reply slip in which they were asked to indicate whether they were willing to take part in the study or not.

Participants' ages ranged from 67 to 83 with a mean age of 74.9. Controls had an average of 17.9 years of full-time education, and were all currently retired. Half the participants had professional or managerial backgrounds, while the remainder had pursued technical or caring occupations. One of the participants was taking anti-depressant medication and four were taking medication for high-blood pressure and cholesterol.

2.2 Materials

Due to the heterogeneity of symptoms that PSP patients can suffer and the differences in progression of PSP, not all patients were able to take all tests and some

tests had to be modified to suit participants' needs (*see* Table 3 for frequency of PSP symptoms in patients) (*see* table 24 in Appendix A for individual symptoms and progression).

Table 3. Frequency of clinical signs in PSP patients in this study at testing time (n= 10).

Symptom	Frequency of patients with this symptom (%)
Supranuclear gaze palsy	100
Unsteady gait and falls	100
Axial dystonia	70
Rigidity	70
Dysarthria	80
Akinesia	60
Focal dystonia	50
Dysphagia	30
Eyelid abnormality	30

Note. Information gathered through the open interview with the patients and their caregivers.

Since the aim of this study is to investigate whether PSP patients are aware of what specific deficits they have, the tests and subtests chosen include difficult visuospatial tasks (e.g. the Line Orientation test) and executive tasks (e.g. the verbal fluency subtest from the ACE), which patients, carers and volunteers should find difficult and easy visuospatial tasks (e.g. dot counting task) which all three groups should find easy and easy executive tasks (e.g. the cognitive estimates test) which volunteers and carers should find easy, but not patients (*see* Table 4).

Table 4. Classification of hard and easy executive and visuospatial tests for the three groups

	Neuropsychological tests	
	Executive tests	Visuospatial tests
Easy for P, C & V	Cognitive Estimates (BADS)	Dot counting (ACE)
Difficult for P,C & V	Verbal fluency (ACE)	Line orientation (BORB)

Note. P = PSP patients, C = carers, V = volunteers

2.2.1 Screening tests

Three short screening tests for cognitive functioning were administered throughout the session.

The Adenbrooke's Cognitive Examination Revised (ACE-R) (Mioshi, Dawson, Mitchell, Arnold, and Hodges, 2006)

This test is used as a general measure of cognitive function, since this test has proven sensitive to detect cognitive deficits in atypical parkinosnian syndromes, for instance PSP (Bak, Rogers, Crawford, Hearn, Mathuranath, and Hodges, 2004). The ACE-R (Mioshi et al., 2006) consists of six subtests: orientation, attention, verbal fluency, language, memory and visuospatial function. The 'orientation' subtest includes fives questions concerning temporal judgements (e.g. 'what day of the week/ month/ year/ and season are we on?') as well as five questions concerning spatial location (e.g. 'what floor/ building/ city /county/ country are you in?'). The 'attention' subtest includes mental calculations (e.g. taking seven away from one hundred) and mental registration of three words and a name and address. In the 'verbal fluency'

component, participants have to give as many words as they can starting with a particular letter in one minute and as many words in the same category (e.g. animals) as possible in one minute. The 'language' subtest requires participants to name 12 pictures, follow some written and verbal commands, repeat single words and sentences and read a set of regular and irregular words. Finally, in the 'visuospatial' component of the ACE-R participants are required to copy two pentagons, a wired cube and to draw a clock face with the numbers and the arms pointing at ten past five.

The Hospital Anxiety and Depression Scale (HADS) (Zigmond and Snaith, 1983)

This test is used as an indication of anxiety and depression levels in participants. It includes 14 questions (half of which are related to depressive symptoms and the other half to anxiety symptoms) which all able participants were asked to complete on their own. Participants with difficulties reading were read out the questions and were asked to indicate with their hand the most appropriate response.

The Perceived Stress Scale (Cohen, Kamarck and Mermelstein, 1983)

The PSS is a measure of global perception of stress during the previous month. It consists of 14 questions which participants have to answer by indicating the most appropriate response in a -point Likert-type scale (O=never to 4 = very often)

2.2.2 Neuropsychological measures

Each participant underwent a comprehensive neuropsychological examination which included tests on memory, executive functions, visuospatial functions and attention.

Memory

Verbal short-term memory was assessed using the Digit Span subtest from the 'Wechsler Memory Scale (WMS III)' (Wechsler et al., 1998). The Digit Span tests participants' ability to remember increasingly longer sequences of digits both forward and in reverse.

Verbal recall was assessed using a word list which contained 15 unrelated words, which participants had to listen to and recall.

Visuospatial short-term memory was assessed using the Corsi Block Tapping Test (Corsi, 1972). In this test, participants were asked to tap an increasingly longer sequence of blocks both forwards and backwards.

The Doors subtest (visual recognition) from the 'Doors and People Test' (Baddeley, Emslie and Nimmo-Smith, 1994). In this subtest, participants were presented with 12 pictures of doors and later asked to identify what door they had previously seen from a set of 4 doors.

Executive Functions Tests

The Cognitive Estimates subtest from the 'Behavioural Assessment of the Dysexecutive Syndrome (BADS)' (Wilson, 1996). In this test, participants were asked four questions regarding common place event and asked to estimate how long each action would take (e.g. 'How long does it take to blow up a party balloon?').

Visuospatial Test

The Orientation Match subtest from the 'Birmingham Object Recognition Battery (BORB)' (Riddoch and Humphreys, 1993). This subtest required participants to decide whether two lines in thirty sets were in the same angle, or parallel, or whether the two lines were in different angles, thus non-parallel.

Attention and Orientation Test

The Elevator task and the Elevator Task with distraction from the 'Test of Everyday Attention (TEA)'(McAnespie, 2001) were used as measures of participants' ability to concentrate and attend to a series of tones. The Elevator subtest was composed of seven sets of tones and required participants to listen and count the number of tones (all the same tone) in a set. The Elevator task with distraction subtest included ten sets of tones and required participants to listen to a set of tones, which included high tones and low tones, and count the number of low tones and ignore the high tones.

2.2.3 Awareness measures

Three types of awareness were measured: emergent awareness, anticipator awareness and metacognitive awareness. Participants were given insight rating scales for the Verbal Fluency and the Dot-Counting subtests (ACE), the Doors subtest (Doors and People Test), the Line Orientation Subtest (BORB), the Cognitive Estimates subtest (BADS), the Digit Span (WMS III), the Corsi Block subtest and the Verbal Recall test.

Table 5 (below) shows the questions that PSP patients, carers and controls were asked before and after each test

Table 5. Insight questions for each group

PSP patients, controls and carers	Question 1: How good do you think is your general ability in this area? Question 2: Now that you know a bit more about the test, how good do you think you will perform on the test? Question 3: Now that you have done the test, how well do you think you did on the test? Question 4: How well do you thing an average person your age would do on this test
Carers only	Question 5: How do you think "the patient" will perform on this task? Question 6: How do you think "the patient" will estimate his ability on this task?

Note. The three groups were asked these questions before and after the verbal fluency and dot counting subtests (ACE), the elevator tasks (TEA), the doors test (Doors and People test), the line orientation test (BORB), the digit span test (WMS-III), the cognitive estimates test (BADS), the verbal recall test and the Corsi block tapping test.

Anticipatory Awareness

Participants were asked to predict their performance in a test in a scale from 'very poor' to 'very good', both when given a general overview of what the test was measuring (e.g. 'this test measures your ability to remember things for a short period of time') and when given detailed instructions of what their task was.

Emergent awareness

Participants were asked to estimate their performance in a test in a scale from 'very poor' to 'very good' after doing the test (e.g. how do you think you performed in this task?).

Metacognitive Awareness

Once participants had finished the test, they were asked to rate in a scale from 'very poor' to 'very good') how they thought a healthy average person their age would perform on such a task.

Caregivers were asked two additional questions; they were asked to rate in a scale from ‘very poor’ to ‘very good’ (c) how they thought the patient would perform in the test and (d) how they thought the patient would rate their ability in that test.

2.2.4 Open interview

Only patients’ and caregivers’ interviews were recorded. Control participants were asked demographic questions such as age, education, occupation, health and medications as well as history of illness in the family.

Patients’ and caregivers’ interviews were more thorough (lasting from 20 minutes to one hour) and asked questions regarding the disease (onset, family history, medication, symptoms, severity of the symptoms, order of appearance of the symptoms, diagnosis), the level of awareness of the patient (accidents and hospitalisations, deficit specificity, severity of impairments) as well as some questions regarding mood change and depression. A voice recorder was used at this stage in order to be able to interact with the patients and caregivers more freely.

2.3 Procedure

2.3.1 Patients and caregivers

Patients and their caregivers were recruited through meetings with the PSP Association in Edinburgh, Glasgow and Towcester, through the months of March to July 2008. At those meetings, the study was presented and information sheets with our contact details were given to the patients and their families. Additional information about the study and our contact details was also published in the PSP Association magazine ‘PSP Matters’ in June 2008.

Patients and caregivers were contacted previous to the testing session in order to adapt and modify any tests to suit the patients' abilities. Testing sessions for patients and caregivers were carried in their own homes but separately (in different rooms of the house) from each other. Only the interview at the end of the testing session was carried out with both patient and caregiver together.

At the beginning of the testing session, they were explained the aim of the study and given the chance to ask any questions; they were also informed that their participation was voluntary and that they could drop at any point in the study without giving a reason. Two patients were unable to sign their own consent forms and thus their main caregiver (which was either their husband or wife) gave consent on their behalf.

The order of the tests was as follows: ACE, Elevator Subtest and Elevator Subtest with Distraction (TEA), Line Orientation Subtest (BORB), Doors Subtest (Doors and People Test), HADS, the Digit Span Subtest (WMS III), the Cognitive Estimates Subtest (BADS), the Verbal Recall Test, Corsi Block Test and the PSS. Before and after each test (except the HADS, PSS and the NART) participants were asked to rate how well they thought they would do or how well they thought they had done in that particular test. The length of the testing sessions varied depending on the stage of the disease that the PSP patients were at and their impairments; some patients were able to complete the set of tests in 1h 20', whilst more severe patients required 2h.

At the end of the testing session, patients and caregivers were recorded whilst they were asked some questions regarding the disease, coping strategies and how it has

affected their mood. At this point they were also debriefed and explained the aim of the study as well as encouraged to ask any questions about the study.

2.3.2 Controls

The same set and order of tests was administered to the control group. At the start of the testing session, participants were given an information sheet which explained the aim of the study and allowed to ask any questions before signing a consent form.

However, participants in this group were tested in the department of Psychology of the University of Edinburgh and their interview at the end of the testing session was not recorded.

3. Results

3.1 All group analysis (PSP patients, carers and controls)

3.1.1 Demographic and screening data

The patient's, carer's and control's groups were well matched in terms of age [F (2, 27) = 1.32, p >.05], years of education [F (2, 27) = 2.34, p >.05].

3.1.2 Performance on neuropsychological tests

Due to the heterogeneous presentation of PSP, and despite the efforts of trying to adapt the tests to the patients' abilities, not all patients were able to complete all tests. Thus, the study is composed of 8 out of 10 complete data sets with regard to PSP patients (see Table 22, in Appendix A for individual scores on each test).

A one-way analysis of variance (ANOVA) was used to compare the three groups' performances on all the neuropsychological tests. There were significant differences between all three groups for all the neuropsychological tests, except for the doors test and the attention and memory subtests of the ACE (see Table 6).

Table 6. Mean scores, standard deviations and significance on neuropsychological tests for the patient, carer and control groups

Cognitive functions (SD)	**Neurocognitive tests**	**PSP patients (n=8)**	**Caregivers (n=10)**	**Controls (n=10)**	***F* - value**	***P* value**
General (SD)	ACE total score	81.25(9.05)	90.40(5.38)	95.50(3.31)	12.28	.00 a, b
Memory mean (SD)	ACE memory	82.21(14.67)	89.23(12.27)	94.23(9.29)	2.21	NS
	Digit Span (WMS-III)	46.25(7.22)	55.67(12.38)	66.33(13.56)	6.67	**.00** b
	Corsi Block	38.89(9.86)	43(6.75)	48.33(5.03)	3.93	**.03** b
	Verbal recall	22.96(12.52)	31.99(12.09)	44.00(14.13)	6.32	**.01** b
	Doors test (Doors and People test	40.83(18.61)	57.50(19.02)	58.33(17.57)	2.87	NS
Attention and concentration mean (SD)	ACE attention and orientation	93.75(8.63)	96.11(5.27)	97.78(3.89)	.99	NS
	Elevator task (TEA)	68.24(21.70)	82.94(18.90)	90.59(14.99)	3.68	**.04** b
Executive functions mean (SD)	ACE fluency	60.71(13.77)	74.29(11.76)	89.29(10.78)	12.66	**.00** b, c
	ACE fluency category	4.71(1.5)	5(1.16)	5.90 (1.29)	6.75	**.01**b
	ACE fluency letter	3.88(.99)	5.30 (.68)	6.60(.52)	9.86	**.00** a, b
	Cognitive estimates (BADS)	40(24.15)	65(17.48)	52.50(21.89)	3.43	**.02** a
Visuospatial functions mean (SD)	ACE dot counting	85(21.10)	97.50(7.9)	100(.0)	3.82	**.03** b
	ACE visuospatial	72.66(16.68)	89.38(8.36)	96.25(5.27)	11.31	**.00** a, b
	Line orientation (BORB)	76.33(12.42)	81.67(7.24)	88.00(7.40)	3.92	**.03** b
Language mean (SD)	ACE language	89.90(8.21)	97.30(4.81)	98.07(2.72)	5.85	**.01** a,b

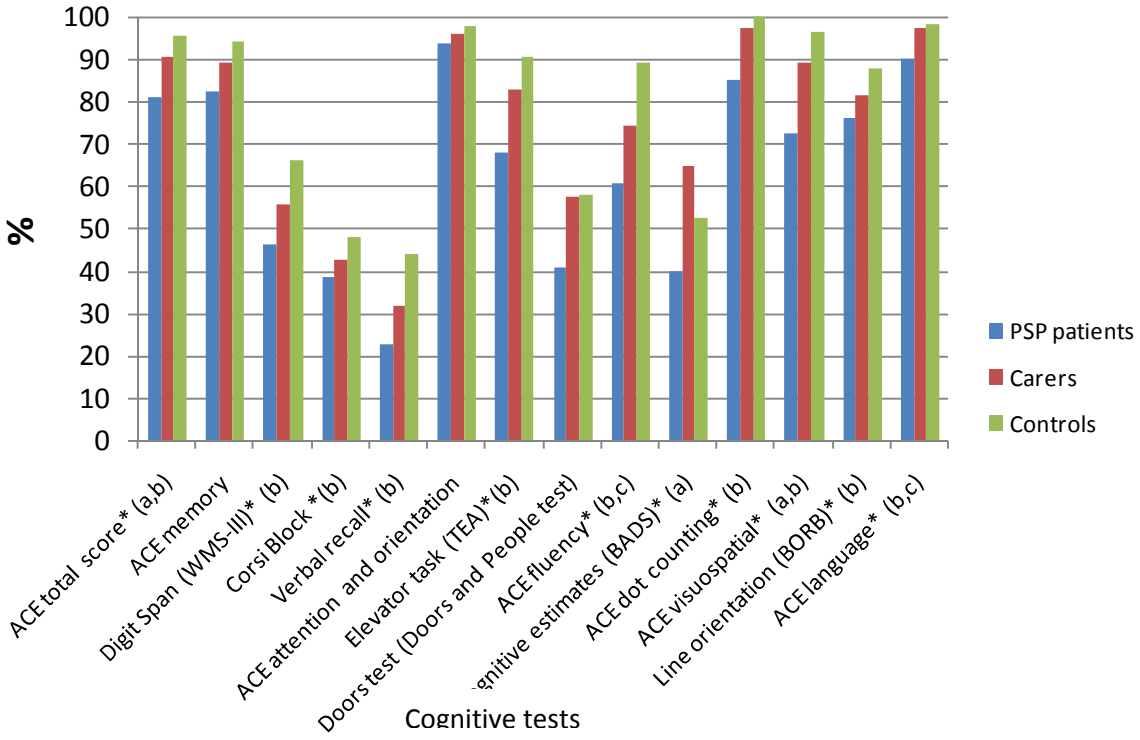
Note. All carers and controls performed above cut-off point for their age on the ACE (85 for the carers and 84 for the control group).The mean performance for patients on the ACE however, was 4 points below their cut-off point. *F*-values in **bold** indicate significant differences between the groups for that particular test. NS = non-significant.

a = post-hoc (Bonferroni) significant differences between patients and carers.
b = post-hoc (Bonferroni) significant differences between patients and controls.
c = post-hoc (Bonferroni) significant differences between controls and carers.

It should be noted that there were significant differences in the dot counting test between patients and controls. It is likely that this is caused by the fact that two patients scored at 50% while the rest performed over 75%. Moreover, consistent with previous literature, there were significant differences between letter and category (animal) fluency between the three groups. Post-hoc tests revealed significant differences between patients and volunteers for the letter fluency and animal fluency subtest (at .05 level); volunteers performed significantly better than patients in these tests. Volunteers also performed significantly better than carers on the category fluency test (at .05 level). No significant differences were found between carers and patients on these two subtests.

As figure 5 shows, controls performed better than carers and patients in all tests except the cognitive estimates test, in which carers performed better (however this difference was not significant). The ANOVA revealed significant differences between the groups' performances in all tests except the doors test and the memory and attention subtests of the ACE.

Figure 5. Mean scores, standard deviations and significance on neuropsychological tests for the patient, carer and control groups



*= significant differences between the three groups
a = post-hoc (Bonferroni) significant differences between patients and carers.
b = post-hoc (Bonferroni) significant differences between patients and controls.
c = post-hoc (Bonferroni) significant differences between controls and carers.

In order to establish that the battery of tests chosen (difficult and easy) was adequate for the study of specificity, comparisons were made within each group, for the visuospatial and executive functions tests (Appendix A, Fig. 17). As we had predicted, all groups performed better at the dot counting test (ACE) than at the line orientation test (BORB). In addition, the three groups performed better at the verbal fluency test (ACE) than at the cognitive estimates test.

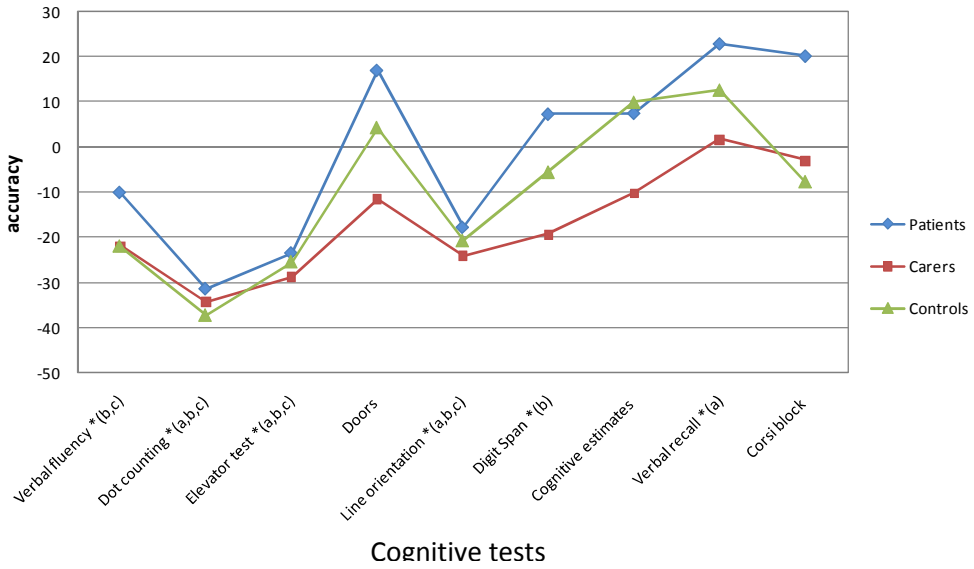
3.1.3 Awareness estimations

Anticipatory awareness

An ANOVA was conducted to examine any significant differences between 'pre-test accuracy' (*i.e.* anticipatory awareness) for all the tests between patients, carers and controls. Pre-test accuracy was calculated by extracting the patients', carers' and controls' actual performance score from their predicted estimated performances once they knew what the test was about (*i.e.* insight question 2).There was a significant difference for the estimated pre-test accuracy on the corsi block test only [F (2, 26) = 3.57, p >.05]. Post-hoc tests revealed that patients were significantly worst at corsi pre-test accuracy in comparison to the control group (at p >.05); patients overestimated their ability on this test (M = 20.11, SD = 30.42) compared to carers and controls (M = -2.90, SD = 22.75 and M = -7.63, SD = 17.93, respectively) (Appendix A, Fig. 16).

Independent samples t-tests were used to compare pre-test accuracy (anticipatory awareness), in patients, carers and controls individually across all tests (*see* Table 7). Patient's t-tests revealed significant differences for the dot counting test, the elevator tasks, the line orientation test and the verbal recall subtest. As figure 6 shows, patients underestimated their abilities on the dot counting test, the elevator tests and the line orientation test whilst they overestimated their ability on the verbal recall test.

Figure 6. Specificity of anticipatory awareness scores on each test for each group



*= significant differences within each group
a = significant differences between tests within the patients' group.
b = significant differences between tests within the carers' group.
c = significant differences between tests within the controls' group

Carer's t-tests also showed significant differences in the verbal fluency test, the dot counting subtest, the elevator tasks, and the line orientation subtest as well as on the digit span subtest, in all of which they underestimated their abilities. Finally, control's t-tests revealed significant differences on the verbal fluency, dot counting, elevator tests and line orientation subtest; in all of which they underestimated their abilities (see Table 7).

Table 7. Specificity of anticipatory awareness scores on each test for each group

	Patients		**Carers**		**Controls**	
Pre-test accuracy on neuropsychological tests	M (SE)	t-value	M(SE)	t-value	M(SE)	t-value
Verbal fluency	-10.08 (12.09)	-.83	-21.89 (5.50)	**-3.98***	-21.89(3.19)	**-6.88****
Dot counting	-31.44 (9.67)	**-3.26***	-34.30 (5.26)	**-6.53****	-37.20(6.21)	**-5.99****
Elevator	-23.53 (9.65**)**	**-2.44***	-28.74 (5.32)	**-5.40****	-25.39(7.61)	**-3.34***
Doors	16.97 (9.87)	1.72	-11.49 (9.36)	-1.23	4.37(5.60)	.78
Line orientation	-17.83 (6.38)	**-2.80***	-23.97 (5.47)	**-4.41***	-20.70(6.56)	**-4.38***
Digit Span	7.25 (9.64)	.75	-19.27 (6.59)	**-2.92***	-5.53(8.55)	-.65
Cognitive estimates	7.44 (10.19)	.73	-10.10 (9.82)	-1.03	10 (7.46)	1.34
Verbal recall	22.82 (7.03)	**3.25***	1.80 (6.62)	.27	12.70(6.95)	1.83
Corsi block	20.11 (10.14)	1.98	-2.90 (7.19)	- .40	-7.64 (5.67)	-1.35

Note. T-values in **bold** indicate significant differences between the anticipatory scores for each test within a group. *significance at p<.05; **significance at p<.001

It should be noted that the three groups had significant differences in three common tests: two visuospatial tests (the dot counting and line orientation) and in an attention test (the elevator task and elevator task with distraction). The three groups underestimated their performances in these three tests.

In order to examine whether patients', carers' and controls' anticipatory awareness estimations were affected by the knowledge of the specific task, a repeated-measures ANOVA was used within each group for each test. Participant's estimations of their general ability in a specific cognitive area (insight question 1) were compared to their predicted estimated performance in the test once they had been explained what the test was about (insight question 2). For patients there was a significant difference

between insight questions 1 and 2 for the verbal fluency subtest [F (1, 7) = 8.405, p <.05]; patients' predicted performance for the verbal fluency test was reduced once they were told what the test was about. Patients' predicted performance for the rest of the tests did not vary significantly once they had been explained the test they were going to do.

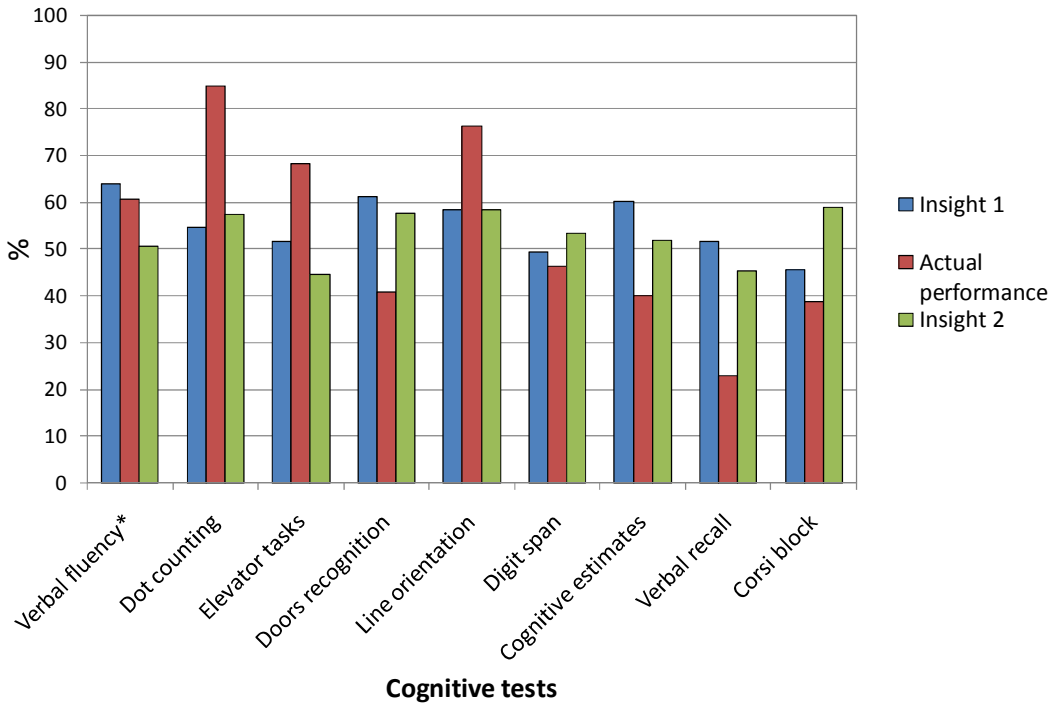
As table 8 and figure 7 show, PSP patients actually underestimated their performances for question 1 on the easy and hard visuospatial tests (the dot counting and line orientation tests, respectively) and on the elevator tests. Interestingly, with the extra information that question 2 included, patients were able to correctly adjust the direction of their predicted performances on five of the nine tests: the verbal fluency, dot counting, doors, cognitive estimates and verbal recall tests.

Table 8. Means and standard deviations for insight estimations 1 and 2 and actual performance for patients in all the neuropsychological tests

	Insight 1	**Actual performance**	**Insight 2**	
Cognitive tests	M (SD)	M (SD)	M (SD)	p-value
Verbal fluency	64(20.16)	60.71(13.77)	50.62(24.27)	<.05*
Dot counting	4.67(24.92)	85(21.10)	57.44(22.56)	NS
Elevator tasks	51.67(16.42)	68.24(21.70)	47.67(18.30)	NS
Doors recognition	61.10(21.22)	40.83(18.61)	57.80(19.48)	NS
Line orientation	58.50(17.45)	76.33(12.42)	58.50(18.28)	NS
Digit span	49.38(12.24)	46.25(7.22)	53.50(23.26)	NS
Cognitive estimates	60.11(18.11)	40(24.15)	51.89(18.89)	NS
Verbal recall	51.67(20.76)	22.96(12.52)	45.22(16.50.35)	NS
Corsi block	45.56(26.35)	38.89(9.86)	59(23.49)	NS

*significant difference between insight 1 and 2. NS = non-significant.

Figure 7. Means for insight estimations 1 and 2 and actual performance for patients in all the neuropsychological tests



*significant difference between insight 1 and 2.

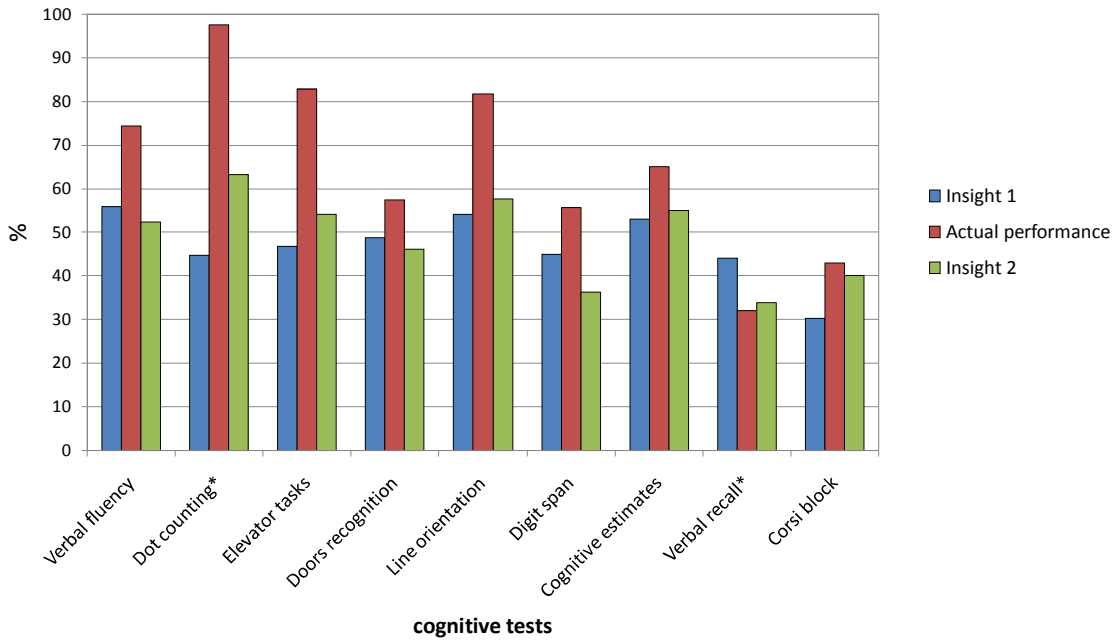
For carers there was a significant difference for the dots subtest [F (1, 9) = 19.29, p<.05] and the verbal recall test [F (1, 9) = 13.35, p<.05]. Whilst carers thought they would do better on the dots subtest once explained what the test was about, the opposite was true for the verbal recall test.

Table 9. Means and standard deviations for insight estimations 1 and 2 and actual performance for carers on all neuropsychological tests

Cognitive tests	Insight 1	Actual performance	Insight 2	
	M(SD)	M (SD)	M (SD)	*p*-value
Verbal fluency	56(19.99)	74.29(11.76)	54.55(14.66)	NS
Dot counting	44.7(12.99)	97.50(7.9)	65(19.34)	<.05*
Elevator tasks	46.80(18.94)	82.94(18.90)	51.45(19.30)	NS
Doors recognition	48.7(21.18)	57.50(19.02)	47.64(18.94)	NS
Line orientation	54.10(17.42)	81.67(7.24)	57.36(20.14)	NS
Digit span	45(17.93)	55.67(12.38)	39.09(23.73)	NS
Cognitive estimates	52.9(24.34)	65(17.48)	56.27(25.12)	NS
Verbal recall	44(20.32)	31.99(12.09)	36(15.61)	<.05*
Corsi block	30.40(19.82)	43(6.75)	43.36(24.82)	NS

*significant difference between insight 1 and 2

Figure 8. Means for insight estimations 1 and 2 and actual performance for carers in all the neuropsychological tests



*significant difference between insight 1 and 2

As shown in table 9 and figure 8, carers underestimate their performance on all tests except on the verbal recall test. They were able however to correctly adjust their prediction on six tests when given extra information (the dot counting, elevator tasks, line orientation, cognitive estimates verbal recall and corsi block tests).

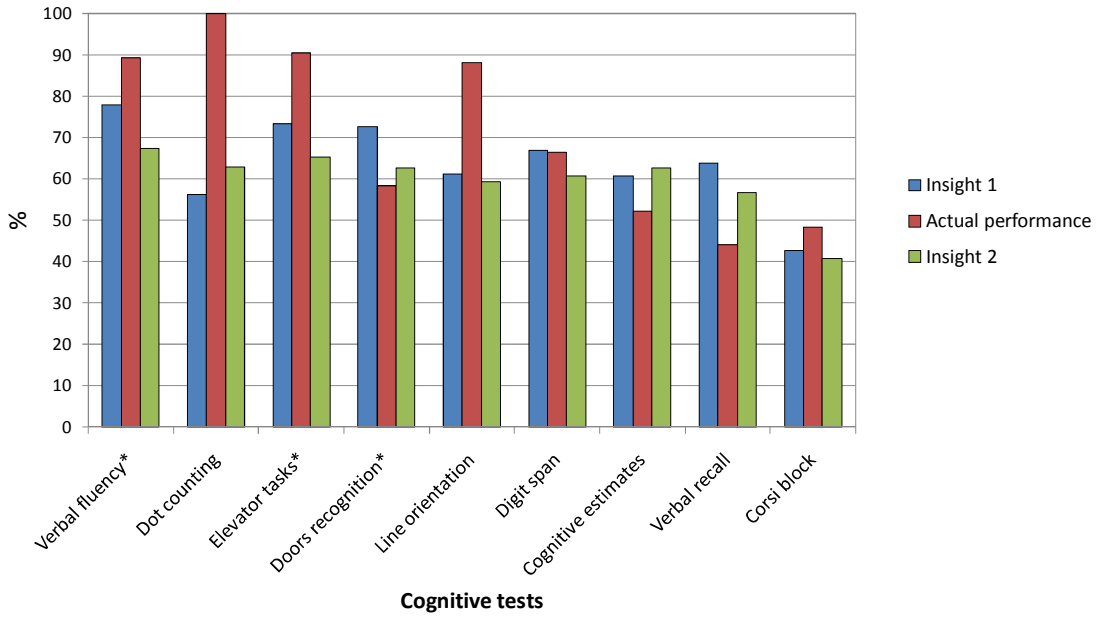
Finally, for controls there were significant differences between insight questions 1 and 2 for the verbal fluency [F (1, 9) = 17.37, p<.05], the elevator tests [F (1, 9) = 5.17, p<.05] and the doors test [F (1, 9) = 5.93, p<.05]. For the three tests, controls lowered their predicted estimations once they knew what the tests were about (see Table 10, Fig. 9).

Table 10. Means and standard deviations for insight estimations 1 and 2 and actual performance for controls on all neuropsychological tests

	Insight 1	**Actual performance**	**Insight 2**	
Cognitive tests	M(SD)	M (SD)	M (SD)	*p*-value
Verbal fluency	77.80(8.25)	89.29(10.78)	67.40(12.83)	<.05*
Dot counting	56.20(23.92)	100	62.80(19.65)	NS
Elevator tasks	73.40(12.62)	90.59(14.98)	65.20(14.82)	<.05*
Doors recognition	72.50(14.58)	58.33(17.57)	62.70(11.90)	<.05*
Line orientation	61.10(18.78)	88(7.40)	59.30(20.47)	NS
Digit span	67(17.26)	66.33(13.56)	60.80(18.33)	NS
Cognitive estimates	60.70(9.37)	52.20(21.89)	62.50(13.39)	NS
Verbal recall	63.80(17.39)	44(14.12)	56.70(16.46)	NS
Corsi block	42.70(20.71)	48.33(5.03)	40.70(20.15)	NS

*significant difference between insight 1 and 2

Figure 9. Means for insight estimations 1 and 2 and actual performance for controls in all the neuropsychological tests



*significant difference between insight 1 and 2

Controls underestimated their performance in five tests (the verbal fluency, dot counting, elevator tests, line orientation, and corsi block tests) and were able to adjust correctly their predictions on four tests (the dot counting, doors, digit span and verbal recall).

Emergent awareness

An ANOVA was conducted to examine significant differences between 'post-test accuracy' (i.e. emergent awareness) for all the tests between patients, carers and patients. Post-test accuracy was calculated by extracting the patients' and carers' actual score from their estimated performances once they had done the test (i.e. insight question 3). There was a significant difference in post-test accuracy for the verbal

fluency subtest [F (2, 25) = 3.71, p<.05]. Post hoc comparisons indicated that in comparison to patients, carers significantly underestimated their performance in the verbal fluency test (at p<.05) (Appendix A, Figure 16).

Independent samples t-tests comparing post-test accuracy (emergent awareness) across tests within the patient group revealed no significant differences in emergent awareness. Only verbal recall approximated significance, which indicates that for patients, emergent awareness is better than anticipatory awareness. Independent samples t-tests were also conducted in order to compare accuracy of emergent awareness in different neuropsychological tests for the carers' and control groups. For carers, there were significant differences on the verbal fluency subtest, the dots subtest, the doors test, the line orientation subtest and the verbal recall test. For controls, there were significant differences on the verbal fluency, dots, elevator tasks, the doors, line orientation and verbal recall tests (Table 11, Fig. 10).

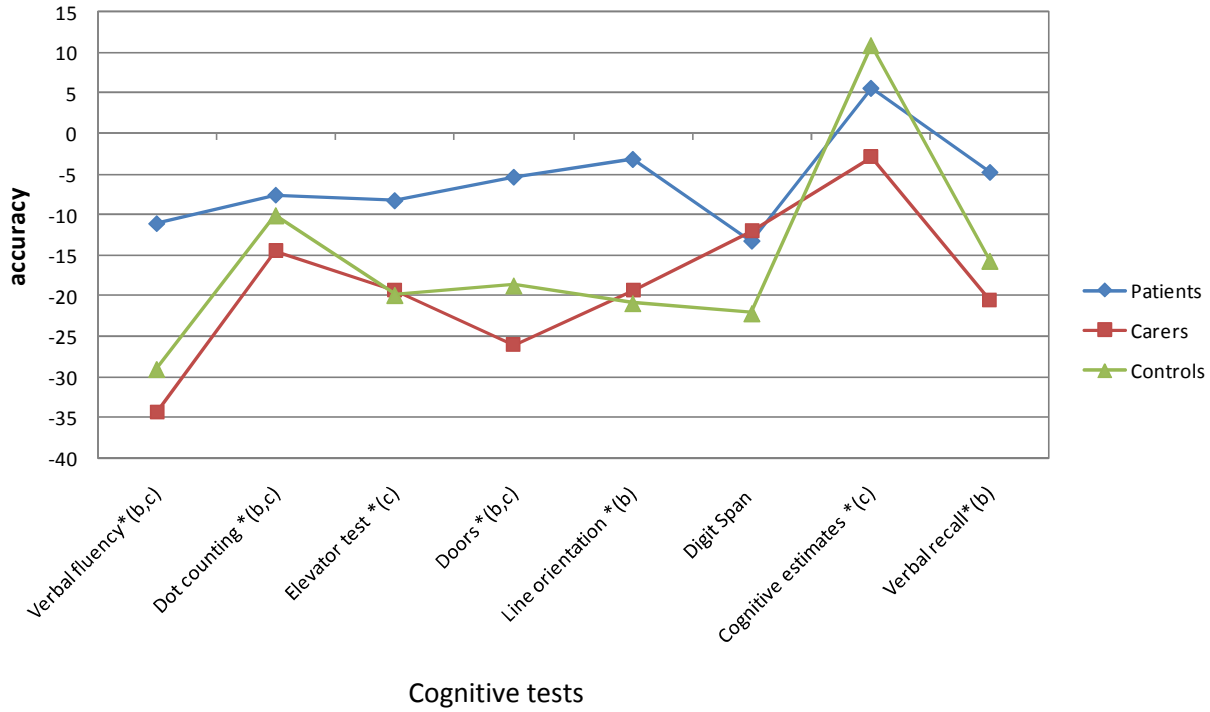
As Table 11 shows, for carers and controls there were significant differences in emergent awareness on three common tests: the verbal fluency, dot counting and doors tests.

Table 11. Specificity of emergent awareness scores on each test for each group

	Patients		**Carers**		**Controls**	
Post-test accuracy on neuropsychological tests	M (SE)	t-value	M(SE)	t-value	M(SE)	t-value
Verbal fluency	-11.09 (7.26)	-1.53	-34.29 (5.57)	**-6.15****	-28.99(5.65)	**-5.13***
Dot counting	-7.56 (8.91)	-.85	-14.40 (3.54)	**-4.06***	-10.10(2.27)	**-4.44***
Elevator	-8.24 (9.19)	-.90	-19.24 (9.69)	**-1.98**	-19.89(5.71)	**-3.48***
Doors	-5.33 (7.61)	-.70	-25.90 (8.62)	**-3.00***	-18.73(5.54)	**-3.38***
Line orientation	-3.13 (4.76)	-.07	-19.27 (7.16)	**-2.69***	-20.90(6.79)	-3.08
Digit Span	-13.25 (6.94)	-1.91	-12.07 (5.42)	-2.23	-22.13(10.06)	-2.20
Cognitive estimates	5.67 (8.90)	-.64	-2.90 (8.45)	-.34	10.80 (8.49)	**1.27***
Verbal recall	-4.74 (4.08)	-1.16	-20.60 (5.44)	**-3.79***	-15.70(5.91)	-2.67
Corsi block	13.55 (8.08)	1.68	5.60 (6.47)	.86	2.81 (5.73)	.50

T-values in **bold** indicate significant differences between the anticipatory scores for each test within a group. *significance at p<.05; **significance at p<.001

Figure 10. Specificity of emergent awareness scores on each test for each group



*= significant differences within each group
a = significant differences between tests within the patients' group.
b = significant differences between tests within the carers' group.
c = significant differences between tests within the controls' group

In order to examine whether patients, carers and controls differed in their awareness estimations before and after doing a test, a repeated-measures ANOVA was conducted comparing pre-test accuracy to post-test accuracy for each participant group. Significant differences were found for patients with PSP in the doors subtest [F (1, 9) = 9.89, p<.05], the dot counting test [F (1, 8) = 15.08, p<.05], and the verbal recall test [F (1, 8) = 18.67, p<.05]; in all of which they showed poorer pre-test accuracy (anticipatory awareness) than post-test accuracy (emergent awareness) (Table 12). The contrary was true for PSP carers; a repeated-measures ANOVA comparing pre-test and post-test accuracy for each test for carers showed significant differences for the verbal fluency test [F (1, 9) = 6.37, p<.05], the doors subtest [F (1, 9) = 7.171, p<.05] and the

verbal recall test [F (1, 9) = 53.228, p<.001], in which carers were less accurate post-test (emergent awareness), whilst they were less accurate in predicting their performance before doing the test for the dot counting subtest (ACE) [F (1, 9) = 24.873, p<.001]. Finally, a comparison of pre-test and post-test means for each test in the control group revealed significant differences for the dot counting test [F (1, 9) = 24.44, p<.05], doors [F (1, 9) = 16.68, p<.05], digit span [F (1, 9) = 5.68, p<.05] and the verbal recall test [F (1, 9) = 21.19, p<.05]. Controls were less accurate at predicting the performance on the dot counting test before doing the test (anticipatory awareness), whilst they were less accurate at emergence awareness estimations for the doors, digit span and verbal recall tests.

Table 12. Means and standard deviations for insight estimations 2 and 3 and actual performance for patients in all the neuropsychological tests

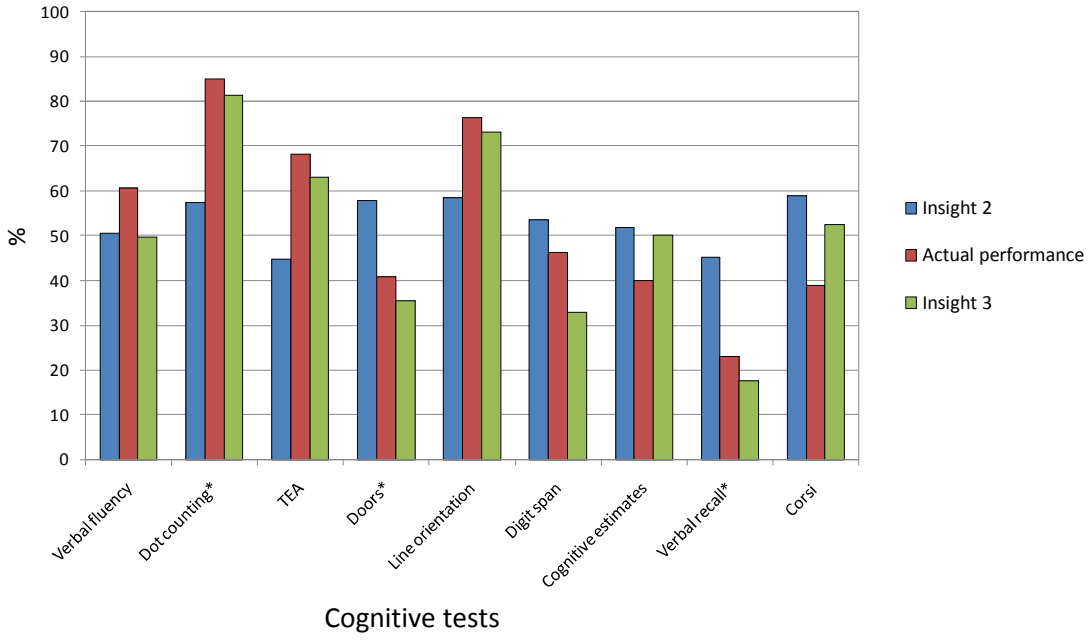
	Insight 2	**Actual performance**	**Insight 3**	
Cognitive tests	M (SD)	M (SD)	M (SD)	*p*-value
Verbal fluency	50.62(24.27)	60.71(13.77)	49.63(11.51)	NS
Dot counting	57.44(22.56)	85(21.10)	81.33(20.59)	<.05
Elevator tasks	47.67(18.30)	68.24(21.70)	63(20.25)	NS
Doors recognition	57.80(19.48)	40.83(18.61)	35.5(19.14)	<.05
Line orientation	58.50(18.28)	76.33(12.42)	73.2(19.29)	NS
Digit span	53.50(23.26)	46.25(7.22)	33(16.78)	NS
Cognitive estimates	51.89(18.89)	40(24.15)	50.11(18.16)	NS
Verbal recall	45.22(16.50.35)	22.96(12.52)	17.67(15.22)	<.05
Corsi block	59(23.49)	38.89(9.86)	52.44(20)	NS

*significant difference between insight 2 and 3. NS = non-significant.

Patients underestimated their performance on four tests before and after doing the tests (the verbal fluency, dot counting, elevator tasks, and line orientation). Except for the verbal fluency, patients correctly adjusted their predictions once having done the

test. On the other tests in which they overestimated their performance before doing them, they correctly managed to adjust their predictions on the correct direction for all tests (Fig.11).

Figure 11. Patients' insight 2 and 3 estimations and actual performance on all tests



* significant differences between insight 2 and 3

Carers underestimated their performance before and after doing the tests for the verbal fluency, dot counting, elevator tasks, doors, line orientation, digit span and cognitive estimates tests, and were able to correctly adjust their predictions on five of those tests (the dot counting, elevator tests, line orientation, digit span and cognitive estimates tests). They were however not able to correctly adjust for the two hard executive tests (the verbal fluency and the doors tests). With regard to the verbal recall

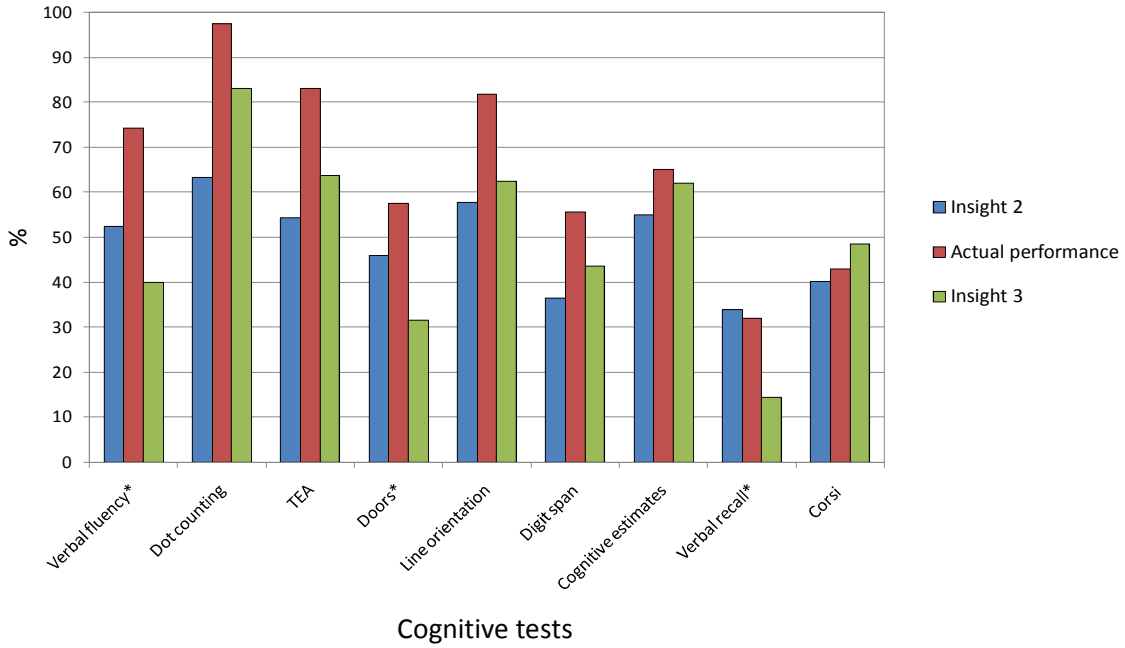
and the corsi block tests, carers overestimated their performances on those tests and correctly adjusted their predictions once having done the tests (Table 13, Fig.12).

Table 13. Means and standard deviations for insight estimations 2 and 3 and actual performance for carers on all neuropsychological tests

Cognitive tests	**Insight 2**	**Actual performance**	**Insight 3**	
	M(SD)	M (SD)	M (SD)	*p*-value
Verbal fluency	54.55(14.66)	74.29(11.76)	40(18.78)	NS
Dot counting	65(19.34)	97.50(7.9)	83.1(13.22)	<.05*
Elevator tasks	51.45(19.30)	82.94(18.90)	63.7(34.29)	NS
Doors recognition	47.64(18.94)	57.50(19.02)	31.6(21.25)	<.05*
Line orientation	57.36(20.14)	81.67(7.24)	62.4(27.27)	NS
Digit span	39.09(23.73)	55.67(12.38)	43.6(23.67)	NS
Cognitive estimates	56.27(25.12)	65(17.48)	62.1(19.73)	NS
Verbal recall	36(15.61)	31.99(12.09)	11.4(11.72)	<.05*
Corsi block	43.36(24.82)	43(6.75)	48.6(18.54)	NS

*significant difference between insight 2 and 3

Figure 12. Carers' insight 2 and 3 estimations and actual performance on all tests



*significant differences between insight 2 and 3

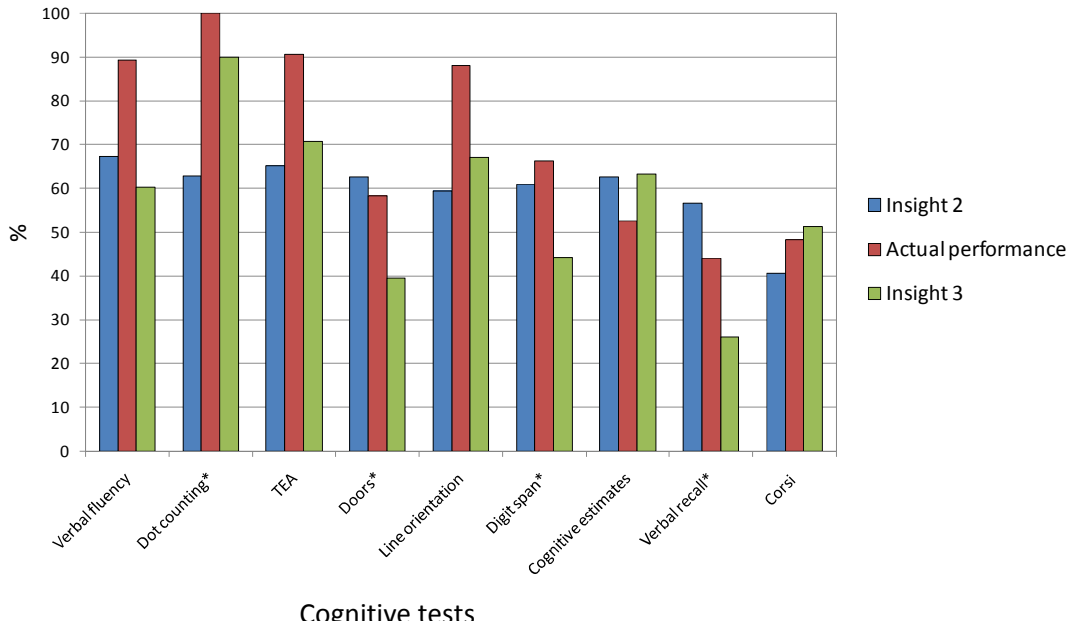
Controls underestimated their performance before doing the tests on the verbal fluency, dot counting, elevator tasks, line orientation, digit span and the corsi block tests, and were also able to correctly adjust their predictions after doing the test on two of those tests (dot counting and elevator tests). They overestimated their performances on the doors, cognitive estimates and verbal recall tests; and were able to correct their estimations on the right direction on two of those tests (the verbal recall and doors tests) (Table 14, Fig.13).

Table 14. Means and standard deviations for insight estimations 2 and 3 and actual performance for controls on all neuropsychological tests

	Insight 2	**Actual performance**	**Insight 3**	
Cognitive tests	M(SD)	M (SD)	M (SD)	*p*-value
Verbal fluency	67.40(12.83)	89.29(10.78)	60.3(18.48)	<.05*
Dot counting	62.80(19.65)	100	89.9(7.19)	NS
Elevator tasks	65.20(14.82)	90.59(14.98)	70.7(17.86)	NS
Doors recognition	62.70(11.90)	58.33(17.57)	39.6(17.58)	<.05*
Line orientation	59.30(20.47)	88(7.40)	67.1(25.60)	NS
Digit span	60.80(18.33)	66.33(13.56)	44.2(24.18)	<.05*
Cognitive estimates	62.50(13.39)	52.20(21.89)	63.3(15.48)	NS
Verbal recall	56.70(16.46)	44(14.12)	26(14.85)	<.05*
Corsi block	40.70(20.15)	48.33(5.03)	51.2(16.54)	NS

*significant difference between insight 2 and 3.

Figure 13. Controls' insight 2 and 3 estimations and actual performance on all tests



*significant differences between insight 2 and 3

Metacognitive awareness

A metacognitive score was computed for patients, carers and controls. For the three groups, the score was calculated by extracting from the groups' mean estimations of others' performances (insight question 4), the mean performance of the volunteers.

A comparison between the three groups revealed a significant difference on the metacognitive scores for the doors test [F (2, 27) = 5.74, p<.05]; post hoc comparisons revealed that patients significantly underestimated others performances' compared to carers and controls.

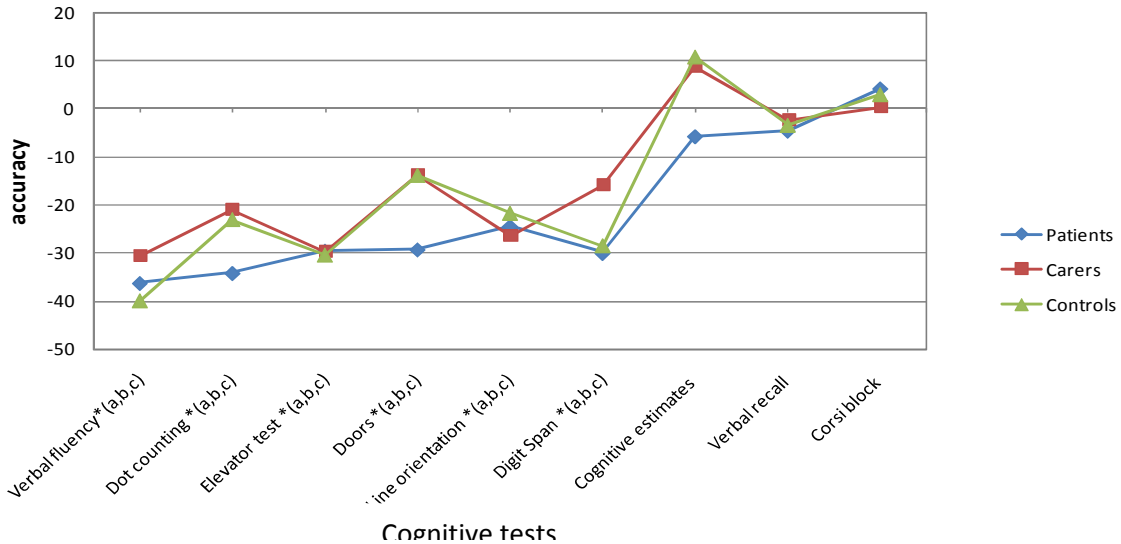
T-tests were used separately on the patient's, carer's and control's metacognitive scores in order to examine test-specificity of metacognitive awareness for each group (Table 15, Fig. 14). Patient's t-tests revealed significant differences for the verbal fluency, dot counting, elevator test, doors test, line orientation test, and digit span test. Carer's t-tests on the metacognitive scores for each test revealed significant differences on the verbal fluency, dot counting, elevator tests, doors test, line orientation test and also the digit span test.

Table 15. Specificity of metacognitive scores on each test with for group

Metacognitive accuracy on neuropsychological tests	**Patients**		**Carers**		**Controls**	
	M (SE)	t-value	M(SE)	t-value	M(SE)	t-value
Verbal fluency	-36.29 (6.75)	**-5.38***	-30.60 (2.22)	**-13.81****	-39.99 (3.18)	**-12.58****
Dot counting	-34.22 (7.67)	**-4.46***	-20.90 (3.13)	**-6.68****	-23.10 (5.99)	**-3.86***
Elevator	-29.59 (4.94)	**-5.98****	-29.79 (4.78)	**-6.23****	-30.49 (7.76)	**-3.93***
Doors	-29.33 (4.36)	**-6.73****	-13.63 (3.99)	**-3.42***	-13.93 (2.70)	**-5.16***
Line orientation	-24.50 (6.31)	**-3.89***	-26.50 (4.24)	**-5.05***	-21.80 (6.06)	**-3.60***
Digit Span	-30.08 (7.15)	**-4.21***	-15.73 (2.87)	**-5.48****	-28.53 (5.79)	**-4.93***
Cognitive estimates	-5.83 (5.31)	-1.09	8.80 (4.76)	.09	10.70 (4.99)	2.15
Verbal recall	-4.67 (6.78)	- .69	-2.30 (2.62)	.40	-3.50 (4.78)	-.73
Corsi block	4.11 (6.69)	.62	.27 (5.86)	.96	2.87 (5.23)	.55

Note. T-values in **bold** indicate significant differences between the anticipatory scores for each test within a group. *significance at p<.05; **significance at p<.001

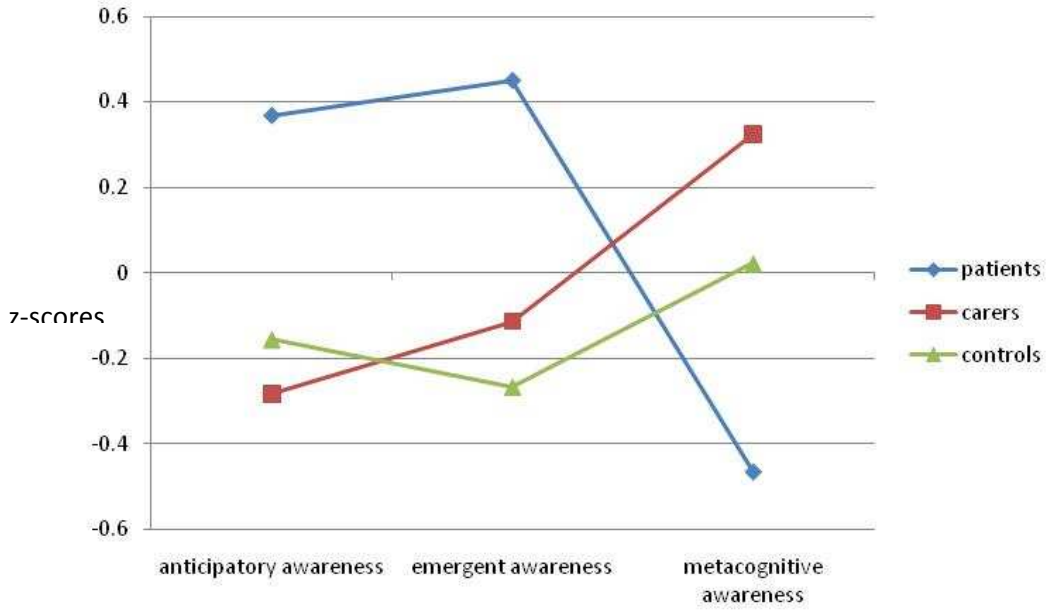
Figure 14. Specificity of metacognitive awareness scores on each test for each group



*= significant differences within each group
a = significant differences between tests within the patients' group.
b = significant differences between tests within the carers' group.
c = significant differences between tests within the controls' group.

Overall anticipatory, emergent and metacognitive awareness scores were computed for each group by summing up all the individual scores for each test. They were then converted onto z-scores and compared between the groups using a one-way ANOVA (Figure 15).

Figure 15. Mean awareness scores (anticipatory, emergent and metacognitive) for each group



Note. comparison between the three awareness measures (z-scores) for the three groups.

3.1.4 Effects of mood on insight estimations and performance

Descriptive statistics for mood, as measured by HADS (anxiety and depression) for all three groups revealed that patients presented with more depressive symptoms and perceived themselves as more stressed than carers did, while carers were more anxious than patients (Table 16). A one-way ANOVA between the three groups revealed significant difference for the HADS-D (HADS depression score); post-hoc contrasts revealed that patients were significantly more depressed than controls (at $p<.05$). An independent samples t-test between carers and patients estimated perceived stress scores (PSS) and mood (HADS anxiety and depression) revealed significant differences only for the PSS score [t (15) = 4.85, p < .001] with patients perceiving themselves as

more stressed [*M*= 31.25, *SE*= 1.80] compared to their carers [*M*= 22.22, *SE*= 1.77] (Appendix A, Table 23 for patients' individual mood scores).

Table 16. Descriptive statistics for mood for PSP patients, their carers and the control group

			HADS Depression	HADS Anxiety	PSS
Group	N	m:f	M (SD)	M(SD)	M(SD)
PSP patients	10	7:3	7.40(2.91)	6.90(4.61)	31.25(5.70)
Caregivers	10	3:7	5.20(3.01)	7.90(3.81)	22.22(5.31)
Controls	10	5:5	2.40(2.46)	4.70(3.92)	NA

Note. M = mean, SD = standard deviation. None of the differences were significant.

Pearson's correlations were performed between HADS –A, HADS-D and PSS with patients', carers' and controls' percentage scores, pre- and post-test accuracy, insight questions 1, 2,3 and the metacognitive score for each cognitive test.

Executive tests

Verbal Fluency test (ACE). There were significant correlations between mood, performance and insight on this test for patients (Table 17).

Table 17. Pearson's correlations between HADS –A, HADS-D and PSS with PSP patients' verbal fluency percentage score, pre- and post-test accuracy, metacognitive scores and insight questions 1, 2 and 3

	VF %	VF pre-test accuracy	VF post-test accuracy	VR metacognitive	VF insight1	VF insight 2	VF insight 3
HADS (A)	.38	**-.81***	**-.71***	-.10	**-.80***	**-.92****	**-.81***
HADS (D)	.64	**-.80***	**-.78***	-.43	-.43	**-.78***	**-.62**
PSS	-.62	.39	.58	.08	.84	.25	.33

*Correlation is significant at the 0.05 level (2-tailed)
**Correlation significant at the 0.01 level (2-tailed)

There were also significant negative correlations for carers between HADS-A and pre-and post-test accuracy (r = -.66 at .05 level, and r = -.66 at .05 level, respectively). Finally, control's performance on the verbal fluency test correlated highly with HADS-A (r = -.93 at .001 level), and their metacognitive awareness on this test correlated negatively with HADS-D (r = -.67, p <.05).

Cognitive estimates test (BADS). Patients' cognitive estimates' pre-test accuracy correlated with HADS-A and HADS-D (r = -.71, p <.05, r = -.81, p <.05, respectively). Insight question 2 for the patients' group also correlated negatively with HADS-A (r = -.72, p <.05). Insight question 3 for the control group also correlated negatively with HADS-D (r = -.65 at .05 level).

Visuospatial tests.

Dot counting test (ACE). There were no significant correlations between mood scores, performance and insight estimations for the patient's group. For carers, there were however, significant negative correlations between HADS-A and HADS-D and

dot counting insight question 2 (r = -.70 at .05, and r = -.85 at .05 respectively). Dot counting post-test accuracy and insight question 3 also correlated positively with PSS (r = .83 at .05, and r = -.75 at .05 respectively).

HADS-A correlated negatively with control's post-test accuracy and insight 3 (r = -.67 at .05 level, and r = -.67 at .05 level, respectively), while HADS-D also correlated negatively with post-test accuracy and insight question 3 (r = -.71 at .05 level, and r = -.71 at .05 level, respectively).

Line orientation test (BORB). For carers, significant Pearson's correlations were present between HADS-D and the line orientation pre-test accuracy (r = -.77 at .01), insight question 1 (r = -.78 at .01) and insight question 2 (r = -.79 at .01) as well as between PSS and pre-test accuracy (r = .67 at .05) and insight question 3 (r = .73 at .05).

Attention test.

Elevator tests (TEA).Patients' insight question 4 and metacognitive awareness in this test correlated negatively with both HADS-A (r = -.89, p <.05; and r = -.89, p <.001 respectively) and HADS-D (r = -.69 p <.05; and r = -.69, p <.05 respectively). For carers, there was a moderate negative correlation between HADS-D and the elevator tasks estimated performance as measured by insight question 3 (r= -0.77 at .01).

Memory tests

Digit span test (WMS-III). Patients' digit span insight question 3 and post test accuracy correlated highly with PSS (r = .78 p <.05and r = -.90 p <.001). Furthermore, for patients there were significant negative Pearson's correlations between HADS-D

and digit span post-test accuracy (r = -.78 at .001) insight question 3 (r = -.78 at .001). Controls' insight 1 estimations also correlated highly with HADS-D (r = -.65 at .001 level).

Verbal recall test. For carers, there were significant correlations between mood and verbal recall percentage, pre-and-post test accuracy and insight questions 1 and 2 (Table 18).

Table 18. Pearson's correlations between HADS –A, HADS-D and PSS with carers' verbal recall percentage score, pre- and post-test accuracy, metacognitive scores and insight questions 1, 2 and 3

	VR %	VR pre-test accuracy	VR post-test accuracy	VR metacognitive	VR insight1	VR insight 2	VR insight 3
HADS (A)	**.70***	**-.65***	**-.70***	-.08	-.32	-.35	-.31
HADS (D)	**.78****	**-.76***	**-.80****	-.40	-.51	-.45	-.37
PSS	.01	.61	.28	.59	**.73***	**.67***	.27

*Correlation is significant at the 0.05 level (2-tailed)
**Correlation significant at the 0.01 level (2-tailed)

Corsi block test. Pearson correlations between the corsi block tapping test performance and carers' mood revealed significant negative correlations between HADS-A and HADS- D with corsi post-test accuracy(r = -.72 at .05, and r= -.66 at .05 respectively).

Doors test (Doors and People test). For patients, there were significant negative correlations between the doors test insight question 1 and the HADS –A (r = -.671, p <.05). The doors subtest scores and insight were also shown to correlate highly with carers' mood (Table 19).

Table 19. Pearson's correlations between HADS –A, HADS-D and PSS scores with carers' doors test percentage score, pre- and post-test accuracy, metacognitive scores and insight questions 1, 2 and 3

	Doors %	Doors pre-test accuracy	Doors post-test accuracy	Doors metacognitive	Doors insight 1	Doors insight 2	Doors insight 3
HADS (A)	**.66***	**-.75***	**-.82****	.13	-.21	-.51	-.46
HADS (D)	**.76***	**-.83****	**-.73***	.33	-.25	-.52	-.25
PSS	.20	.41	.18	.34	.61	**.69***	.32

*Correlation is significant at the 0.05 level (2-tailed)
**Correlation significant at the 0.01 level (2-tailed)

Finally, depression in the control group correlated highly with their estimated performance (Question 1) (r = -.90, p <.001).

3.2 Patients vs. carers

3.1.1 Demographic and screening data

Patients and carers' groups were well matched in terms of age [t (18) = .50, p >.05], years of education [t (18) = .08, p >.05], as well as on depression (HADS-D) [t (18) = 1.67, p >.05], and anxiety (HADS-A) [t (18) = -.53, p >.05]. However, significant differences existed in their perceived stress as measured by the PSS questionnaire [t (15) = 4.85, p < .001], with patients perceiving themselves as more stressed [M= 34.50, SE= 1.80] compared to their carers [M= 22.22, SE= 1.77].

3.1.2 Performance on neuropsychological tests

An ANCOVA between patients' and carers' performances on each test, once accounted for their differences in their PSS scores, revealed no significant differences in their performance for any of the cognitive tests (Table 20).

Table 20. Mean scores and standard deviations on neuropsychological tests for the patients' and carers' groups

Cognitive functions	Neurocognitive tests	PSP patients (n=8)	Caregivers (n=10)	*F* - value	*P* value
General (SD)	ACE total score	82.14 (9.39)	80.22 (5.67)	2.62	NS
Memory mean (SD)	ACE memory	82.21(14.67)	89.23(12.27)	.33	NS
	Digit Span (WMS-III)	46.25(7.22)	55.67(12.38)	1.84	NS
	Corsi Block	38.89(9.86)	43(6.75)	.29	NS
	Verbal recall	22.96(12.52)	31.99(12.09)	1.14	NS
	Doors test (Doors and People test)	40.83(18.61)	57.50(19.02)	.46	NS
Attention and concentration mean (SD)	ACE attention and orientation	93.75(8.63)	96.11(5.27)	.98	NS
	Elevator task (TEA)	68.24(21.70)	82.94(18.90)	.68	NS
Executive functions mean (SD)	ACE fluency	57.14(13.77)	74.29(11.76)	3.49	NS
	Cognitive estimates (BADS)	40(24.15)	65(17.48)	4.58	NS
Visuospatial functions mean (SD)	ACE dot counting	85(21.10)	97.50(7.9)	1.30	NS
	ACE visuospatial	72.66(16.68)	89.38(8.36)	2.64	NS
	Line orientation (BORB)	76.33(12.42)	81.67(7.24)	.61	NS
Language mean (SD)	ACE language	89.90(8.21)	97.30(4.81)	2.79	NS

Note. All carers performed above cut-off point for all tests. P-values were non-significant for all tests

3.1.3 Awareness estimations

An ANCOVA including PSS scores as covariates revealed no significant differences in any of the tests between patients and carers for any of the awareness types (anticipatory, emergent or metacognitive).

Pearson's correlations were then performed between the carers' estimations of the patients' abilities on a task (insight question 5), and the patients' actual performance on that task. Carers estimations correlated highly only with the patients' own performance for the dot counting test *(r* = 0.67, *p*< .05), meaning that carers estimations of patients performance were not accurate for most of the cognitive tests.

Pearsons's correlations were also performed between carers' predictions of patients own performance estimations for each test (insight question 6) and patients own estimations pre-and post-test (patients insight question 1, 2, and 3). There was one moderately significant correlation only for the cognitive estimates test between question 6 and insight (*r* = 0.72, *p*<.05).

3.3 Patients vs. volunteers

3.2.1 Demographic and screening data

Patients and volunteers' groups were well matched in terms of age [*t* (18) = -.95, *p* >.05], years of education [*t* (18) = -1.76, *p* >.05] and anxiety (HADS-A) [*t* (18) = 1.15, *p* >.05]. Significant differences existed between depression (HADS-D) [*t* (18) = 4.15, *p* <.001] patients were more depressed than volunteers (*M* = 7.40, *SE* = .92 for patients vs. *M* = 2.40, *SE* = .78).

3.2.2 Performance on neuropsychological tests

An ANCOVA between patients and the control group's performance revealed significant difference in the digit span, corsi block test, verbal recall, verbal fluency, and the visuospatial and language subtests of the ACE, as well as the ACE total score (Table 21).

Table 21. Mean scores and standard deviations on neuropsychological tests for the patients' and control groups

Cognitive functions	Neurocognitive tests	PSP patients (n=8)	Controls (n=10)	*F* – value	*P* value
General (SD)	ACE total score	81.25(9.05)	95.50(3.31)	19.13	.001
Memory mean (SD)	ACE memory	82.21(14.67)	94.23(9.29)	4.33	NS
	Digit Span (WMS-III)	46.67(7.70)	66.33(13.56)	7.37	.00
	Corsi Block	42.38(5.68)	48.33(5.03)	6.39	.02
	Verbal recall	20.95(12.42)	44(14.13)	.57	.00
	Doors test (Doors and People test)	48.81(16.27)	58.33(17.57)	.04	NS
Attention and concentration mean (SD)	ACE attention and orientation	93.75(8.63)	97.78(3.89)	1.92	NS
	Elevator task (TEA)	75.63(14.51)	90.59(14.99)	2.10	NS
Executive functions mean (SD)	ACE fluency	62.24(14.12)	89.29(10.78)	18.31	.00
	Cognitive estimates (BADS)	42.86(18.90)	52.50(21.89)	.34	NS
Visuospatial functions mean (SD)	ACE dot counting	85.71(19.67)	100	1.74	NS
	ACE visuospatial	72.66(16.69)	96.25(5.27)	7.55	.00
	Line orientation (BORB)	75.24(14.51)	88(7.40)	1.02	NS
Language mean (SD)	ACE language	89.90(8.21)	98.08(2.72)	5.16	.01

Note. All carers performed above cut-off point for all tests. P-values in **bold** indicate significant differences between the groups for that particular test.

3.2.3 Awareness estimations

Anticipatory awareness

An ANCOVA including HADS-D scores as covariates was used to compare pre-test accuracy (anticipatory awareness), in patients, carers and controls individually

across all tests. The results revealed no significant differences in any of the tests between patients and carers.

Emergent awareness

An ANCOVA was conducted to examine significant differences between 'post-test accuracy' (i.e. emergent awareness) for all the tests between patients and controls (including HADS-D scores as covariates). The analysis revealed a significant difference in the verbal fluency post-test accuracy between patients and controls [F (1, 14) = 8.06, p >.05], in shich carers significantly overestimated their performance (M = 67.40, SD = 12.83) compared to patients (M = 50.62, SD = 24.27).

Metacognitive Awareness

An ANCOVA including HADS-D scores as covariates on control's and patient's metacognitive scores revealed a significant difference in the verbal fluency test [t (1,14) = 10.75, p>.05]; where volunteers underestimate significantly others' performances on this test by 39.99%.

3.3 Volunteers vs. carers

3.3.1 Demographic and screening data

The carers and volunteers groups were well matched in terms of age [t (18) = -1.65, p >.05], years of education [t (18) = -1.95, p >.05] and anxiety (HADS-A) [t (18) = 1.85, p >.05], however, there were significant differences between HADS-D [t (18) = 2.28, p <.05]; carers were more depressed than controls (M = 5.20, SE = .95 for carers vs. M = 2.40, SE = .78 for controls).

3.3.2 Performance on neuropsychological tests

An ANCOVA revealed significant differences between carers and the control group on the verbal fluency test [F (1, 16) = 17.14, p >.05]' the verbal recall [F (1, 16) = 6.05, p >.05] and the corsi block tests [F (1, 16) = 9.87, p >.05]. Controls performed significantly better on all those tests compared to the carers group.

3.3.3 Awareness estimations

Anticipatory awareness

An ANCOVA including HADS-D as a covariate indicated that there were no significant differences between 'pre-test accuracy' (i.e. anticipatory awareness) for all the tests between carers and controls.

Emergent awareness

An ANCOVA including HADS-D as a covariate indicated that there were no significant differences between 'post-test accuracy' (i.e. emergent awareness) for all the tests between carers and controls.

Metacognitive awareness

An ANCOVA including HADS-D as a covariate revealed one significant difference for the verbal fluency metacognitive score [F (1, 16) = 5.27, p>.05]; volunteers underestimated others' performances significantly more than carers did (M = -39.99, SD = 10.06 for volunteers vs. M = -30.69, SD = 7.03 for carers).

4. Discussion

Performance and cognitive awareness were assessed in a group of patients with PSP, their main carers and a group of controls. The battery of tests used measured five main cognitive areas: memory, attention and concentration, language, visuospatial and executive function abilities. Within the visuospatial and executive function tests included, there were both hard and easy to perform tests. These were purposely added in order to examine specificity of cognitive awareness among the three groups.

A comparison of the performance between the three groups revealed significant differences in all the tests (the digit span, corsi block, elevator tasks, cognitive estimates, line orientation and the verbal recall test) with the exception of the doors test. Patients were also significantly impaired on most of the subtests of the ACE, except for the attention and memory subtests.

This pattern of results is consistent with those found by Bak and colleagues (2005b), and is in keeping with our current understanding of the cognitive abilities and the distribution of pathological changes in patients with PSP. As Bak et al. (2005b) suggest, the significant verbal fluency impairment on the ACE subtest is likely to be caused by pathological changes to the basal ganglia and reflect the subcortical 'core deficit' proposed by Albert et al (1974). The impairments present in the language and visuospatial subtests of the ACE however are more likely to reflect a more cortical pathology, involving more frontal and fronto-parietal structures instead.

Despite not finding a significant impairment in patients on the attention and orientation subtests of the ACE, our results revealed an impaired performance of patients on the Test of Everyday Attention (TEA) subtests.

These results can be explained by the fact that all our patients presented ocular motor dysfunction. The relationship between motor ocular dysfunction and cognitive abilities was examined by Esmonde, Gils, Gibson and Hodges (1996). In their study, they used a battery of tests similar to that used in this study, which also included the elevator subtests of the TEA, and were able to demonstrate a significant correlation between oculor motor dysfunction and performance on the TEA. The more severe the ocular motor dysfunction was, the worse their performance on the TEA.

Interestingly, our patients were also impaired on the dot counting subtest of the ACE. However, these results can be explained by two main factors: a significantly impaired performance by only two patients on this test (thus causing our normally distributed scores to skew) and chance. The dot counting task was specifically included as an easy visuospatial task, which patients should not find difficult. This test consists of four squares with dots in them. Participants are asked to visually count the number of dots in each square. Two cases in this study were only able to count the number of dots in two of the four squares correctly, while the majority were able to count all the squares. Therefore, the present impairment on this task may be the result of those two significantly impaired cases distorting the final average. Their impaired performance on this task may be caused by other factors such as impaired vision or stimuli presentation.

There is however another factor which is highly related to the low scores found in this test, that is the phenomenon of chance. Compared to other tests, there are only four questions in the dot counting test. An error in one of the questions will reduce an individual's performance by 25%; thus, taking account the ocular difficulties that some patients with PSP have, it is possible that they were not able to clearly see the stimuli and miscounted. This would cause their performance to decrease by 25% each time.

Future research that takes into account the layout of the tests as well as oculo-motor dysfunction in PSP should shed light on this matter.

Moreover, with regard to the performance of patients on the cognitive tests, and consistent also with previous literature, patients were better at recognition (the doors test) than at recall (the verbal recall test) (Haist, Shimamura and Squire, 1992). Despite carers and controls performing better than patients on the doors test, this difference was not significant.

Finally, the performance displayed by the three groups on the hard and easy visuospatial and executive tests, was consistent with the predicted performance for these tasks. The three groups performed better at the easy visuospatial test (the dot counting test) than at the hard test (the line orientation test). The verbal fluency test was harder than the cognitive estimates test for all groups.

From the pattern of results obtained, we can conclude that the battery of tests used was appropriate for this study.

Insight was then measured in the context of a multidimensional model which distinguishes between three types of awareness: anticipatory, emergent and metacognitive awareness (Toglia and Kirk, 2000).

Anticipatory awareness was measured by asking participants to predict what they thought their performance would be before doing a test. Emergent awareness was measured in a similar way to anticipatory awareness, by asking participants to rate what they thought their performance was after doing a test. Metacognitive awareness instead, required participants to rate how they thought another person of their own age would do in the same test. Each awareness type was then examined individually across groups and tests.

Following O'Keefe et al (2007) suggestion that mood may influence predictive accuracy, this study also took into account depression and anxiety (as measured by the HADS questionnaire) on the three groups, as well as perceived stress levels (as measured by the PSS questionnaire) on both carers and patients' performance estimations.

Anticipatory awareness

A comparison between the three groups (PSP patients, carers and controls) revealed that patients were significantly worse than controls at predicting their performance on the corsi block test. All three groups considerably underestimated their performance on three common tests: the easy and hard visuospatial tests (the dot counting and line orientation tests) and the elevator subtests. Whereas carers and controls tended to significantly underestimate their predicted performance for all tests, patients underestimated their performance only on half the tests (mainly the two visuospatial tests and the attention subtests of the TEA). In fact, they had a tendency to overestimate their performance, especially on the executive tasks (which they were meant to find difficult). Other studies on anticipatory awareness in PSP patients have also found this pattern of results (O'Keeffe et al, 2007 on PSP patients; and Souchay et al, 2003 on FTD patients).

O'Keeffe and colleagues (2007) proposed that PSP patients' inaccuracies in predicting what their performance would be like are likely to be due to the frontal atrophy or frontal deafferentation present in those patients. This suggestion was based on their finding that performance on the Frontal Assessment Battery test (FAB) was a strong predictor of anticipatory awareness accuracy. Although this study used different

frontal tests to that of O'Keefe (2007), it also found that PSP patients were impaired on frontal lobe tests, which gives further support to the frontal atrophy hypothesis.

We then examined whether the amount of information that a participant was given had an influence on their predicted estimations. Patients were able to use the extra information and adjust their predictions accordingly for most tests, with the exception of the corsi block test and the digit span test. This however does not mean that they were accurate in their predictions. Instead, it means that they were able to process and use the extra information given in a correct manner.

The level of information given before each test across studies can vary enormously and as this results indicate, PSP patients require more detailed information (possibly with examples) in order to correctly predict their accuracy in the test. Thus, for comparison purposes, it seems necessary that studies measuring awareness should agree on the level of information given to a participant prior to doing a test.

The type of test and level of information given seemed to have little effect on caregivers' estimations of their own performances. Carers underestimated their performance on all tests before and after given extra information about the test. Nevertheless, they were able to appropriately adjust their estimations on five of the nine tests (the dot counting, line orientation, cognitive estimates and corsi block tests).

The issue of inaccurate estimations by carers on their own performance is in line with Clare, Wilson, Carter, Roth and Hodges (2002) findings on early stage AD patients and their carer's. They found that carers were not accurate predictors of either patients' performance or their own performance. In fact, they underestimated both. We could argue that these results suggest a '*caring for progressive supranuclear palsy patient's effect'*, in that caring for those patients makes people more aware of their own deficits

and thus more prone to underestimate their own performance. Maylor, Smith, Della Sala and Logie (2000) found a somewhat similar effect on carers of AD patients, although on the opposite direction. In their study, AD carers overestimated their own performance compared to an age-matched group of controls and termed this effect '*caring for Alzheimer's Disease patient's effect*'. Maylor et al (2000) proposed that carers overestimation of their own performances was due to a comparison they made between their abilities and those of the patients they are caring for, causing them to have an inflated opinion of their abilities.

This issue was then further investigated, and carers' estimations of patients' performances as well as their estimations of patients' estimations were compared to the patients' actual performance and patients' actual estimations. There were only moderate correlations for two of the tests, which supports the idea that carers' estimations are not very accurate. This result is important since it highlights the methodological issue of using carers as controls to give reliable and accurate estimations and validate rating scales (Maylor, et al., 2000).

In contrast, controls underestimated their performance before given extra information on five tests only (the verbal fluency, dot counting, elevator tests, line orientation and corsi block tests); whilst they overestimated on the rest. Extra information on the test prior to testing meant that controls were able to adjust their predictions on four tests only.

Emergent awareness

The ability to retrospectively estimate one's cognitive abilities has been linked to the anterior cingulated cortex and the dorsolateral prefrontal regions (Hester, Foxe,

Molholm, Shapner and Garavan, 2005). This ability however, has also been linked to sustained attention deficits in frontal lobe damage (McAvinue, O'Keefe, McMackin and Robertson, 2005).

Due to the frontal atrophy and impaired performance on the TEA subtests, we would expect PSP patients to be significantly impaired on this type of awareness. Our results however show a different pattern of performance than predicted. Patients were able to accurately estimate their performances once they had done the test. In fact, their estimations were more accurate than those done by their carer's and control group.

The carer and control groups had a tendency to underestimate their performances for all tests; only the corsi block test was an exception on all three groups. Patients instead, tended to overestimate their performances after testing on most of the tests.

Furthermore, carers and controls also seemed to be particularly bad at estimating their performance on the easy and hard visuospatial and executive functions tests compared to the rest of the tests (the attention and memory tests).

A comparison was then made between pre-and post-test accuracy for the three groups, which revealed that patients underestimated their performance prior to test on four tests (the verbal fluency, dot counting, line orientation and elevator subtests), in most of which they managed to adjust their predictions correctly once they had done the tests. They overestimated their performance prior to the tests on the remaining five tests, and were then able to correctly adjust the direction of their predictions once they had done the tests on all of those tests. Patients, however, seemed to have difficulties adjusting their performance on the executive tests.

It thus seems that PSP patients are able to correctly use the information gathered about their performance by doing the task and apply it to their estimations of

performance. However, if we compare their predictions before and after the performance of the tests to that of carers and controls, it is possible to suggest that PSP patients are accurate on their predictions because they are overestimating their performance. Contrary to carers and controls, and consistent with the pathology present in PSP, we could hypothesise that patients' disinhibition causes them to overestimate their performance, while the control and carer's groups are afraid to be perceived as immodest and thus underestimate their performances (Fernandez-Duque and Black, 2007). Inhibition is a cognitive skill which heavily relies on the executive system. Although several studies support the view that executive functions rely on the anterior and posterior cortical regions of the brain (*e.g.* Fuster, 2000), most studies agree that executive functions rely heavily on the integrity of the frontal lobes (Souchay et al., 2003). The executive system however does not only encompass this function, it also includes other functions such as monitoring one's recent and past performances, generating future goals or even altering behavioural patterns in response to feedback (Souchay et al., 2003; Roberts and Pennington, 1996).

As shown by comparing pre-and post-test accuracy scores, patients are able to use feedback information from their performance on the test to modify their predictions correctly, thus suggesting that the frontal pathology present in PSP patients does not affect the executive system as a whole. Instead, it seems that PSP presents with a pattern of spared and impaired frontal 'sub-functions' and not a generalised executive impairment. Similar findings to those found in PSP patients, have been found in normal ageing studies. Connor, Dunlosky and Hertzog (1997) found an increase in accuracy from before to after the study in normal ageing adults. They suggested that this might be due to the fact that after study predictions involve exposure to the task prior to

estimating one's performance, thus providing participants with information about the test procedure and an opportunity to self-monitor.

Interestingly, carers and controls were able to adjust correctly their estimations post-test for most tests. They however had difficulties with the difficult executive functions tasks (the verbal fluency) and on the doors test. Although the doors test is a recognition memory test, it could be argued that this test has an executive component. Performance on these two tests seems to be harder to judge for healthy adults and thus specificity of cognitive domain seems to be related to prediction accuracy.

In summary, our results show that patients are more accurate at their predictions pos-test (emergent awareness) than pre-test (anticipatory awareness). These results are consistent with those found by O'Keeffe and colleagues (2007). They also found that PSP patients had better emergent awareness than anticipatory and metacognitive awareness. However, awareness in their study, despite being conceptually identical to the one in this study, was measured in a different manner. O'Keefe et al (2007) used different tests to measure different types of awareness in three groups of patients (PSP, CBD and FTD patients). It is possible to suggest that the methodology used does not allow for comparisons to be made between the different awareness types since, as this study suggests, there are differences in the accuracy of PSP patients' estimations depending on what cognitive domain the tests examine.

Metacognitive awareness

Metacognitive awareness refers to one's ability to estimate others' performances. Some psychologists have argued that metacognitive judgement is secondary to actual performance (Kruger and Dunning, 1999; Fernandez-Duque and Black, 2007). In line

with this hypothesis, we could suggest that the knowledge and skills that are required to perform a test and predict one's own abilities in one domain, are also required for judging other's abilities in the same domain. Therefore, poor pre-and post-test prediction accuracy is also likely to cause poor metacognitive accuracy.

Our results showed that the three groups had a tendency to underestimate others' abilities. In particular, the three groups significantly underestimated others' performances on 6 tests: the verbal fluency, the dot counting, the elevator tests, the doors test, the line orientation test and the digit span test. These results show that people are not only likely to underestimate others' performances on tests they find difficult, but they also seem to underestimate their performance for tests which they find very easy (*e.g*. the dot counting test). Although the three groups were inaccurate at predicting how others would perform, our results reveal that carers and controls were more accurate than patients at those estimations.

A comparison between the three types of awareness in this study has revealed that patients have a tendency to overestimate their own performance pre-and post-test. They however have a tendency to underestimate others' performances on the same tests. Carers on the other hand seem to underestimate their performances both pre-and post test and overestimate others' performances on the same tests. Finally, controls show a similar pattern of results to that of carers, i.e. they underestimate their performance, but they seem to be more accurate at judging how other people their age will do.

These results seem to be in agreement with the clinically observed unawareness of personality and cognitive changes which many of those patients display, as well as with findings suggesting that patients with frontal lobe damage, such as fronto-temporal dementia patients, have a tendency to overpredict their performance compared to other

patients, such as AD patients (Souchay et al, 2003). Furthermore, it should be noted that the relationship between an individual's actual performance on a test and their estimation of their performance may reflect a statistical artefact. Since subjective estimates of performance are never perfectly correlated with one's actual performance, judgements of performance will have a tendency to regress towards the mean. This means that participants who perform worst at a given domain will have a tendency to overestimate their performances on that domain, while those who perform best at a particular domain will tend to underestimate their performance on that specific domain (Fernandez-Duque and Black, 2007).

Despite these biases on the beliefs each individual has on one's own performance being the norm for most of the population; neurological and psychiatric disorders, including PSP, have a tendency to exacerbate those biases (Fernandez-Duque and Black, 2007).

Mood

While it is recognised that depression is a common symptom in PSP (Albert, Feldman and Willis, 1974; Chiu, 1995) the evidence that depression has an influence on cognitive abilities and insight remains inconclusive (Sevush and Leve, 1993; Feher et al, 1991; Reed et al, 1993; De Bettignes et al, 1990; Verhey et al, 1993, Esmonde et al., 1996).

Depression and anxiety, as defined by the scores on the HADS, were frequently present (only two cases scored below cut-off point of 7 on depression, while 5 cases scored below cut-off point of 7 on anxiety) in the patients in our study. Most carers exhibited depressive symptoms also (6 cases scored above cut-off point of 7 on

depressions), while only three cases scored highly on anxiety. In contrast, only three controls scored highly on depression and one case scored highly on anxiety. A comparison of the mood scores between the three groups revealed that patients were the most depressed group followed closely by their caregivers. Furthermore, the patient group perceived themselves as more stressed compared to the carers' group.

There were some significant correlations between mood, performance and insight estimations for the three groups on the cognitive tests, especially with the hard executive function tests (the verbal fluency) as well as the doors test.

Consistent with the anxiety and depression scores found for the control group, correlations between awareness estimations and mood for controls were low or non-significant.

Anxiety and depression correlated strongly with anticipatory and emergent awareness estimations for patients on the verbal fluency, digit span and cognitive estimates tests. For carers, anxiety and depression correlated highly with anticipatory and emergent awareness estimations on the doors, verbal fluency, dot counting, line orientation and verbal recall tests. Metacognitive awareness was only correlated with patient's estimations on the elevator tasks. These correlations suggest that the more depressed the patients were, the more they underestimated theirs or others performance for specific tests.

In agreement with Esmonde and colleagues' (1996) results, performance on any of the neuropsychological tests and mood were not highly correlated, which gives further support to the suggestion that mood and neuropsychological factors are independent from each other (Esmonde et al, 1996).

In summary, it seems that patients with PSP have a specific pattern of awareness with not only general differences between emergent, anticipatory and metacognitive awareness (emergent being better than anticipatory and metacognitive awareness), but also domain-specific differences across different cognitive areas (with patients making more mistakes in executive tests than in any other types of tests). This suggests that the pattern of awareness in those patients may be caused by an impairment to a particular function believed to rely on the frontal lobes, for instance disinhibition or attention and concentration.

These results also show that the estimations made by the primary carers on the patients' as well as on their own performance are inaccurate. This finding is very interesting from a methodological point of view, and adds concerns to studies which rely solely on the estimations made by the carers.

Clinical implications

Progressive Supranuclear Palsy is a degenerative disease with motor and cognitive impairments. This means that as the disease progresses, patients are forced to depend entirely on family and friends to perform everyday functions. However, carers are often ill-equipped to deal with the emotional and physical demands that this disease places on them, thus increasing the caregiver burden. As mentioned in the introduction, caregiver burden encompasses the physical, psychological or emotional, social and financial problems that can be experienced by family members caring for impaired elderly adults (George and Gwyther, 1996).

An important factor which has been related to negative aspects of rehabilitation outcome as well as increasing the caregiver burden is the patient's awareness of their

own deficits. Thus study of disordered awareness has very important clinical implications. For instance, in patients with traumatic brain injury (TBI), lack of awareness has been found to correlate negatively with vocational and residential status, (Trudel, Tryon and Purdum, 1998) as well as with poor compliance to medication (McGlynn and Shacter, 1989; Cotrell and Wild, 1999).

It is important also to mention that studies have found that as diseases such as Alzhemeir's disease progress, awareness impairments also have a tendency to increase (Sevush and Leve, 1993), which can lead to more frustration in the carers and may even lead to some hospitalisations and accidents. From talking to the patients and caregivers in this study, it was clear that PSP patients had a tendency to overestimate their abilities, and consequently they would carry out tasks that some of them were not capable of achieving alone (*e.g.* trying to get up and walk to the toilet, or make themselves a cup of tea).

Moreover, depression and behavioural disturbances are also common symptoms in PSP which can have an effect on awareness and be extremely unsettling for carers (Fernandez-Duque and Black, 2007).

If we take into account all these factors and their effect on the patient's and carer's quality of life, it is clear that the study of insight is a priority and should be taken into account in any rehabilitation programme and family counselling.

As this study has shown, and consistent with the results found by O'Keefe et al (2007), lack of awareness is a common symptom in PSP patients. However the awareness impairment that these patients have is not a generalised impairment, it depends on the 'object of insight' (Markova and Berrios, 2001). Patients with PSP are

able to process information correctly and adjust their predictions for their own performance estimations. Thus, their emergent awareness (post-test accuracy) seems to be better than their anticipatory awareness (pre-test accuracy). However, they are not able to estimate accurately the performances of others (metacognitive awareness). Their awareness deficits seem to also be sensitive to the cognitive domain which is being assessed. As this study has shown they seem to be worst at estimating their performances on difficult than on easy tests, as well as on those tests they are bound to find difficult due to their cognitive impairments (in this case executive tests). These results have important implications not only because they can help improve the quality of life of the patients and their carers and help with possible diagnosis and rehabilitation programmes or compensatory mechanisms, but also as they highlight the importance of selecting an appropriate battery of tests depending on the population studied, defining and characterising the complex concept of awareness and the methodology used to measure it. These issues will now be reviewed further.

Misdiagnosis in PSP is common among patients and can lead to frustration in the patients and clinician. In depth studies on the different symptoms that patients with this disease show will allow clinicians to be able to diagnose the disease sooner, and decrease the patient's feeling of frustration and concern. Earlier diagnosis will mean that patients seek for support and information earlier which in turn will help them understand the symptoms associated with this disease, especially the behavioural and emotional symptoms.

An in depth understanding of the deficits in awareness that PSP patients may have can not only help with diagnosis but also can also help clinicians and the carers with

possible coping techniques which can reduce the accidents and hospitalisation that these patients suffer.

PSP patients have shown that they are able to correctly adjust their estimations once they have done a task or given extra information and examples of what the test involves, which in turn suggests that they are able to process extra information. This has both practical and theoretical implications in that patients may benefit from being explained why the can or cannot do a task in detail and using examples. It also suggests that deficits in awareness in patients are very complex and should be assessed using a multidimensional approach which distinguishes between self- and others-awareness (the awareness an individual has of his/her own performance and the awareness an individual has of other people's performances) as well as between cognitive and functional domains.

Furthermore, by talking to the PSP patients in this study, it is possible to suggest that some differences exist between the level of cognitive and physical awareness in those patients. Future studies should examine this hypothesis by including a measure of physical severity as well as questions regarding physical impairments.

The correlations found between mood and awareness estimations on the different cognitive tests reflect the need to investigate this phenomenon further. Anxiety and depression in the patients in this study have been shown to correlate highly with awareness measures of executive functioning. This indicates that mood may have a significant influence on the patients' predictions of what their performance is. Mood is therefore important in the study of awareness and should be taken into account in any

study which measures an individual's ability to accurately predict his/her own performance.

Finally, carers in this study scored significantly high on anxiety and seemed to have a biased opinion of what the patients or themselves were able to do. It is possible that these inaccuracies were caused by anxiety and depression levels in this particular carer group, as correlations between mood and awareness estimations show; however, it is unlikely to find carers of cognitive or physically impaired patients who do not show a degree of anxiety and depression. It is vital that future studies using carers as accurate estimators of patients performances also take into account mood and distress levels.

This is the first study that investigates the specificity of cognitive awareness in PSP patients, their carers and a group of controls, and as we have seen the results obtained have very important theoretical and practical implications. Future studies investigating awareness impairments in this disease should account for this specificity and further examine how these deficits impact everyday tasks and the quality of life of both patients and their carers.

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6. Appendix A: Tables and figures

Table 22. individual scores of PSP patients on neuropsychological tests

Cognitive functions	Neurocognitive tests	PSP patients									
		1	2	3	4	5	6	7	8	9	10
General performance	ACE total	NA	75	96	74	NA	78	74	80	95	78
Memory (%)	ACE memory	NA	56.79	100	76.92	NA	88.46	88.46	73.08	100	73.08
	Digit Span (WAIS)	NA	43.33	53.33	36.67	NA	40	43.33	56.67	53.33	43.33
	Corsi Block	NA	36.67	53.33	40	16.67	40	36.67	40	46.67	40
	Verbal recall	40	13.33	13.33	6.67	NA	26.67	20	13.37	33.33	40
	Doors test (Doors and People test)	16.67	33.33	41.67	75	25	41.67	25	66.67	50	33.33
Attention and concentration (%)	ACE attention and orientation	NA	100	100	94.44	NA	94.44	77.78	100	100	83.33
	Elevator tasks (TEA)	41.18	47.06	94.12	94.12	47.06	70.59	64.71	100	70.59	52.94
Executive functions (%)	ACE fluency	NA	35.71	71.43	57.14	NA	64.29	21.43	71.43	78.57	75
	Cognitive estimates (BADS)	0	25	50	25	75	25	25	50	75	50
Visuospatial functions (%)	ACE dot counting	50	75	100	75	100	50	100	100	100	100
	ACE visuospatial	NA	81.25	100	50	NA	56.25	68.75	62.50	87.5	75
	Line orientation (BORB)	76.67	70	73.33	46.67	73.33	86.67	86.67	90	83.33	76.67
Language (%)	ACE language	NA	92.31	100	80.77	NA	76.29	88.46	88.46	100	92.31

Note. F-value and P-values before controlling for premorbid intelligence (NART), depression (HADS), years of full-time education and age.

Table 23. individual scores of PSP patients on mood tests

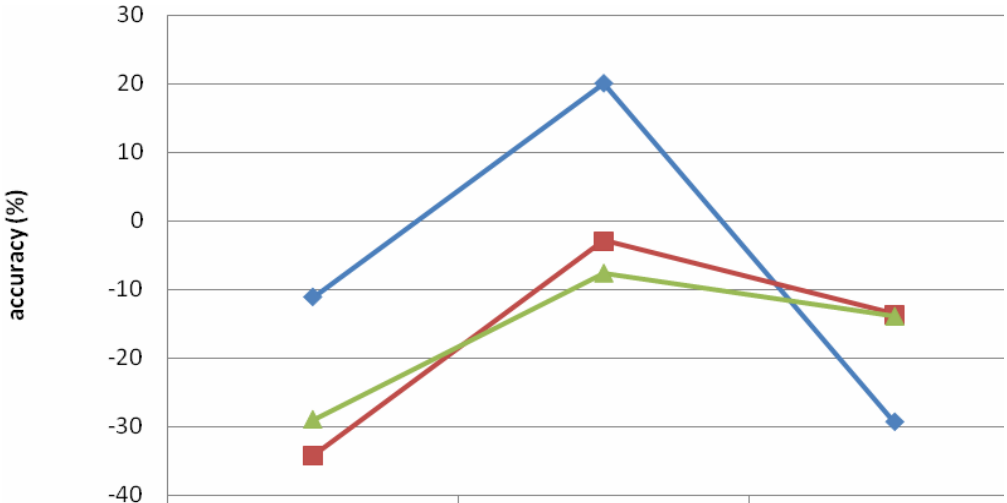
Mood Tests	**PSP patients**									
	1	2	3	4	5	6	7	8	9	10
HADS –Anxiety (max.21)	7	5	2	10	13	11	1	5	13	2
HADS – Depression (max.21)	7	5	7	7	13	10	2	7	9	7
PSS (max.56)	NA	NA	33	39	33	32	30	30	17	34

Note. Cut off points for both HADS-A and HADS-D is at 7 points.

Table 24. Individual progression of PSP symptoms

Patients	Year diagnosed	Progression of symptoms (in order from first appearance as carers and patients remember)
1(KC)	2002	Unsteady gait and falls, akinesia, focal dystonia, supranuclear gaze palsy, axial dystonia, rigidity eyelid abnormality, dysarthria, dysphagia
2(IA)	2004	Unsteady gait and falls, dysarthria, supranuclear gaze palsy, akinesia
3(JW)	2002	Unsteady gait and falls, dysphagia, dysarthria, supranuclear gaze palsy, rigidity
4(DH)	2002	Unsteady gait and falls, supranuclear gaze palsy, rigidity, akinesia, focal dystonia, axial dystonia, dysarthria
5(PC)	2004	Unsteady gait and falls, rigidity, axial dystonia, dysarthria, dysphagia, supranuclear gaze palsy, eyelid abnormality
6(RM)	2006	Unsteady gait and falls, axial dystonia, rigidity, dysarthria, supranuclear gaze palsy
7(RO)	2005	Unsteady gait and falls, akinesia, focal dystonia, axial dystonia, rigidity, supranuclear gaze palsy, dysarthria
8(MP)	2004	Unsteady gait and falls, focal dystonia, axial dystonia, akinesia, supranuclear gaze palsy, dysarthria, eyelid abnormality
9(MC)	2008	Unsteady gait and falls, supranuclear gaze palsy
10(JM)	2006	Unsteady gait and falls, axial dystonia, rigidity, dysarthria, suprannuclear gaze palsy, focal dystonia, akinesia

Figure 16. Significant awareness differences between PSP patients, carers and controls in specific neurological tests



	Verbal fluency emergent awareness	Corsi Anticipatory awareness	Doors metacogntive awareness
patients	-11.09	20.11	-29.33
carers	-34.29	-2.9	-13.63
controls	-28.99	-7.63	-13.93

Figure 17. Comparison between difficult and easy visuospatial and executive test across the three groups